

COLUMBIA LIBRARIES OFFSITE

HEALTH SCIENCES STANDARD



HX00047929

**RECAP**

RC41

Os5

Columbia University 1913  
in the City of New York v.5

College of Physicians and Surgeons *Copy 1*

Library



*Gift of*

*Dr. C.R. Murray*







Digitized by the Internet Archive  
in 2010 with funding from  
Open Knowledge Commons



Tracy | 37 Hillhouse Ave.  
New Haven  
Conn

## CONTRIBUTORS.

BARKER, LEWELLYS F., M.D., LL.D.

BRAMWELL, EDWIN, M.B., F.R.C.P. (Edin. and  
Lond.), F.R.S. (Edin.).

BURR, CHARLES W., M.D.

BUZZARD, E. FARQUHAR, M.D., F.R.C.P. (Lond.).

CLARK, L. PIERCE, M.D.

COLLINS, JOSEPH, M.D.

CUSHING, HARVEY, M.D.

DOCK, GEORGE, M.D.

EMERSON, CHARLES P., M.D.

HOLMES, GORDON M., M.D., M.R.C.P.

JELLIFFE, SMITH ELY, A.M., M.D., Ph.D.

McCARTHY, DANIEL J., M.D.

McCRAE, THOMAS, M.D., F.R.C.P. (Lond.).

RUSSEL, COLIN K., M.D.

SACHS, BERNARD, M.D.

SOUTHARD, E. E., A.M., M.D.

SPILLER, WILLIAM G., M.D.

STEINER, WALTER R., M.D.

TAYLOR, EDWARD W., M.D.

THOMAS, HENRY M., A.M., M.D.

# MODERN MEDICINE

## ITS THEORY AND PRACTICE

IN ORIGINAL CONTRIBUTIONS BY AMERICAN AND  
FOREIGN AUTHORS

EDITED BY

SIR WILLIAM OSLER, BART., M.D., F.R.S.

REGIUS PROFESSOR OF MEDICINE IN OXFORD UNIVERSITY, ENGLAND; HONORARY PROFESSOR OF MEDICINE IN  
THE JOHNS HOPKINS UNIVERSITY, BALTIMORE; FORMERLY PROFESSOR OF CLINICAL MEDICINE IN  
THE UNIVERSITY OF PENNSYLVANIA, PHILADELPHIA, AND OF THE INSTITUTES OF MEDICINE  
IN MC GILL UNIVERSITY, MONTREAL, CANADA

AND

THOMAS McCRAE, M.D.

PROFESSOR OF MEDICINE IN THE JEFFERSON MEDICAL COLLEGE, PHILADELPHIA; FELLOW OF THE ROYAL COLLEGE OF  
PHYSICIANS, LONDON; FORMERLY ASSOCIATE PROFESSOR OF MEDICINE, THE JOHNS HOPKINS UNIVERSITY

VOLUME V

DISEASES OF THE NERVOUS SYSTEM—DISEASES  
OF THE LOCOMOTOR SYSTEM

*SECOND EDITION, THOROUGHLY REVISED*

ILLUSTRATED



LEA & FEBIGER  
PHILADELPHIA AND NEW YORK  
1915

PC 40

025

2.2

1.5

1.5

Entered according to the Act of Congress, in the year 1915, by  
LEA & FEBIGER,  
in the Office of the Librarian of Congress. All rights reserved.



## CONTRIBUTORS.

---

LEWELLYS F. BARKER, M.D., LL.D.,

Professor of Clinical Medicine in the Johns Hopkins University; Visiting Physician to the Johns Hopkins Hospital, Baltimore, Md.

EDWIN BRAMWELL, M.B., F.R.C.P. (EDIN. AND LOND.), F.R.S. (EDIN.),

Assistant Physician to the Royal Infirmary and Lecturer on Medicine in the School of the Royal College, Edinburgh, Scotland.

CHARLES W. BURR, M.D.,

Professor of Mental Diseases in the Medical Department of the University of Pennsylvania, Philadelphia.

E. FARQUHAR BUZZARD, M.D., F.R.C.P. (LOND.),

Out-Patient Physician to the National Hospital for the Paralyzed and Epileptic, and to St. Thomas' Hospital, London; Consulting Neurologist to the Royal Free Hospital, and to the Hospital for Diseases of the Throat, London, England.

L. PIERCE CLARK, M.D.,

Formerly First Assistant Physician to the Craig Colony for Epileptics; Consulting Neurologist to the Manhattan State Hospital; Neurologist to Randall's Island Hospital; Chief Consultant of Letchworth Village for Mental Defectives.

JOSÉPH COLLINS, M.D.,

Physician to the Neurological Institute of New York, New York City.

HARVEY CUSHING, M.D.,

Professor of Surgery in the Harvard University Medical School, Boston, Mass.; Formerly Associate Professor of Surgery in the Johns Hopkins University.

GEORGE DOCK, M.D.,

Professor of Medicine in the Washington University Medical School, St. Louis, Missouri.

CHARLES P. EMERSON, M.D.,

Professor of Medicine in the Indiana University, School of Medicine, Indianapolis, Ind.; Late Associate in Medicine and Resident Physician in the Johns Hopkins University and Hospital; Late Superintendent of the Clifton Springs Sanatorium.

GORDON M. HOLMES, M.D., M.R.C.P.,

Assistant Physician to the National Hospital for the Paralyzed and Epileptic, London; Physician to the Out-Patients, Seamen's Hospital, London, England.

SMITH ELY JELLIFFE, A.M., M.D., PH.D.,

Adjunct Professor of Diseases of the Mind and Nervous System in the Post-Graduate Medical School and Hospital; Visiting Neurologist to the City Hospital; Consulting Neurologist to the Manhattan State Hospital, New York.

DANIEL J. MCCARTHY, M.D.,

Professor of Medical Jurisprudence in the Medical Department of the University of Pennsylvania; Neurologist to the Henry Phipps Institute and to the Philadelphia and St. Agnes' Hospitals, Philadelphia.

THOMAS McCRAE, M.D., F.R.C.P. (LOND.),

Professor of Medicine in the Jefferson Medical College, Philadelphia.

COLIN K. RUSSEL, M.D.,

Lecturer in Clinical Neurology in McGill University, Montreal, Canada

BERNARD SACHS, M.D.,

Alienist and Neurologist to Bellevue Hospital; Neurologist to the Mt. Sinai Hospital; Consulting Physician to the Manhattan State Hospital, New York.

E. E. SOUTHARD, A.M., M.D.,

Bullard Professor of Neuropathology in the Medical School of Harvard University, Boston, Mass.; Director of the Psychopathic Department of the Boston State Hospital; Pathologist to the State Board of Insanity, Massachusetts.

WILLIAM G. SPILLER, M.D.,

Professor of Neuropathology and Associate Professor of Neurology in the Medical Department of the University of Pennsylvania, Philadelphia.

WALTER R. STEINER, M.D.,

Visiting Physician to the Hartford Hospital; Consulting Physician to the Hartford Orphan Asylum; Consulting Physician to the Middlesex Hospital of Middletown, Conn.

EDWARD W. TAYLOR, M.D.,

Assistant Professor of Neurology in the Medical School of Harvard University, Boston, Mass.; Chief of the Neurological Service in the Massachusetts General Hospital, Boston, Mass.

HENRY M. THOMAS, A.M., M.D.,

Clinical Professor of Neurology in the Johns Hopkins University; Neurologist to the Johns Hopkins Hospital and Dispensary, Baltimore, Md.

# CONTENTS.

## PART I.

### DISEASES OF THE NERVOUS SYSTEM.

#### CHAPTER I.

INTRODUCTION TO DISEASES OF THE NERVOUS SYSTEM . . .	17
By LEWELLYS F. BARKER, M.D.	

#### CHAPTER II.

DISEASES OF THE MOTOR TRACTS . . . . .	80
By WILLIAM G. SPILLER, M.D.	

#### CHAPTER III.

THE COMBINED SYSTEM DISEASES OF THE SPINAL CORD . . .	125
By COLIN K. RUSSEL, M.D.	

#### CHAPTER IV.

SCLEROSIS OF THE BRAIN . . . . .	140
By EDWIN BRAMWELL, M.B., F.R.C.P. (EDIN. AND LOND.)	

#### CHAPTER V.

THE DISEASES OF THE MENINGES . . . . .	162
By EDWIN BRAMWELL, M.B., F.R.C.P. (LOND. AND EDIN.)	

#### CHAPTER VI.

DIFFUSE AND FOCAL DISEASES OF THE SPINAL CORD . . .	178
By E. FARQUHAR BUZZARD, M.D., F.R.C.P. (LOND.)	

#### CHAPTER VII.

TOPICAL DIAGNOSIS OF DISEASE OF THE BRAIN . . . . .	265
By JOSEPH COLLINS, M.D.	

#### CHAPTER VIII.

APHASIA . . . . .	292
By JOSEPH COLLINS, M.D.	

#### CHAPTER IX.

TUMORS OF THE BRAIN AND MENINGES . . . . .	308
By HARVEY CUSHING, M.D.	

#### CHAPTER X.

HYDROCEPHALUS . . . . .	351
By HARVEY CUSHING, M.D.	

## CHAPTER XI.

- ACUTE ENCEPHALITIS AND BRAIN ABSCESS . . . . . 359  
 BY E. E. SOUTHARD, M.D.

## CHAPTER XII.

- DISEASES OF THE CEREBRAL BLOODVESSELS . . . . . 387  
 BY HENRY M. THOMAS, A.M., M.D.

## CHAPTER XIII.

- DISEASES OF THE CEREBRAL NERVES . . . . . 471  
 BY E. W. TAYLOR, M.D.

## CHAPTER XIV.

- DISEASES OF THE PERIPHERAL NERVES . . . . . 533  
 BY GORDON M. HOLMES, M.D., M.R.C.P.

## CHAPTER XV.

- EPILEPSY . . . . . 592  
 BY L. PIERCE CLARK, M.D.

## CHAPTER XVI.

- NEURASTHENIA. THE TRAUMATIC NEUROSES AND PSYCHOSES 624  
 BY CHARLES W. BURR, M.D.

## CHAPTER XVII.

- HYSTERIA . . . . . 654  
 BY SMITH ELY JELLIFFE, A.M., M.D.

## CHAPTER XVIII.

- MIGRAINES. NEURALGIA. PROFESSIONAL SPASMS. OCCUPA-  
 TION NEUROSES. TETANY . . . . . 709  
 BY SMITH ELY JELLIFFE, A.M., M.D.

## CHAPTER XIX.

- PARALYSIS AGITANS. CHOREA. CHOREIFORM AFFECTIONS.  
 INFANTILE CONVULSIONS . . . . . 764  
 BY DANIEL J. MCCARTHY, M.D.

## CHAPTER XX.

- MYASTHENIA GRAVIS. PARAMYOCLONUS MULTIPLEX. PERI-  
 ODIC PARALYSIS . . . . . 798  
 BY DANIEL J. MCCARTHY, M.D.

## CHAPTER XXI.

- ASTASIA-ABASIA. ADIPOSIS DOLOROSA . . . . . 815  
 BY DANIEL J. MCCARTHY, M.D.

## CHAPTER XXII.

- SYPHILITIC DISEASES OF THE CENTRAL NERVOUS SYSTEM . . 822  
 BY BERNARD SACHS, M.D.

## CHAPTER XXIII.

- AMAUROTIC FAMILY IDIOCY (TAY-SACHS DISEASE) . . . . 863  
 BY BERNARD SACHS, M.D.

## PART II.

## DISEASES OF THE LOCOMOTOR SYSTEM.

## CHAPTER XXIV.

MYOSITIS—MYOTONIA CONGENITA: THOMSEN'S DISEASE. . . .	871
By WALTER R. STEINER, M.D.	

## CHAPTER XXV.

ARTHRITIS DEFORMANS . . . . .	895
By THOMAS McCRAE, M.D., F.R.C.P. (LOND.)	

## CHAPTER XXVI.

OSTEOMALACIA . . . . .	947
By GEORGE DOCK, M.D.	

## CHAPTER XXVII.

ACHONDROPLASIA. HYPERTROPHIC PULMONARY OSTEO- ARTHROPATHY. PAGET'S DISEASE. OSTEOGENESIS IM- PERFECTA. OSTEOPSATHYROSIS. LEONTIASIS OSSEA. MICROCEPHALUS. FACIAL HEMIATROPHY . . . . .	955
By CHARLES P. EMERSON, M.D.	

INDEX . . . . .	993
GENERAL INDEX . . . . .	1029



# DISEASES OF THE NERVOUS SYSTEM.

## CHAPTER I.

### INTRODUCTION TO DISEASES OF THE NERVOUS SYSTEM.

By LEWELLYS F. BARKER, M.D., LL.D.

#### THE SENSES AND THEIR SYMPTOMATOLOGY.

It is convenient to divide the senses into two groups, *interior* and *exterior*. The interior senses are those of which we project the sensations into some part of our own bodies. They include pain, movement sense, position sense, resistance sense, the vestibular sense, hunger, thirst, the sexual sense, and the sense of fatigue. The exterior senses are those of which we project the sensations into the world outside ourselves. They include sight, hearing, taste, smell, touch and pressure; and the two temperature senses, heat and cold.

**The Interior Senses.—Pain.**—Physiologists now regard this as a special sense, and not simply as a negative feeling tone associated with overstimulation of other sense organs (von Frey). This pain sense has a punctiform distribution in the skin. As tested by very fine needles, the points are more numerous even than the touch points. The strength of the stimulus required to call forth a sense of pain is called the threshold stimulus, and this varies for different pain points. The localization of pain sensations is due less to the pain nerves than to the simultaneous stimulation of the sense of touch.

Visceral pain is often not localized in the viscera themselves, but is referred to certain areas in the skin (Head). The cutaneous hyperesthesias in cardiac, gastric, intestinal, and prostatic diseases are examples of such mis-reference. The pain sense of the skin and viscera has been designated by Head and Rivers as *protopathic* sensibility, in contrast with touch and temperature senses, which they call *epicritic* sensibility.

Sometimes the peripheral nerves are oversensitive to pressure, particularly at places where they emerge from bony openings (*Valleix's points*).

**Deep or Mesoblastic Sensibility. Sensations of Movement of Parts of the Body; of Their Position, and of Resistance. Bathyesthesia.**—Under the term "muscle sense," a whole series of different sensations have been roughly lumped together. This is due largely to the lack of sharpness of these sensations in consciousness. We must distinguish at least the following: (1) The sense of position of the individual parts of the body;

(2) the sense of movements of these parts; (3) sensations of resistance and weight; (4) the stereognostic sense; (5) the so-called vibration sense.

1. *Sense of Position of Individual Parts of the Body.*—The normal person is able actively to imitate with one-half of his body positions in which portions of the other half are passively placed.

2. *Sense of Movement of Individual Parts of the Body.*—When the eyes are closed and active or passive movements of a part of the body are made, a normal person is able to state (a) that a movement has occurred, (b) the degree of this movement, and (c) its direction. This sense of movement differs from the sense of position. In normal individuals any movement that the examiner himself can see should be recognized. When a joint has to be flexed or extended through several degrees before a sensation is detected, the movement sense is subacute. It should be remembered that smaller movements can be recognized in the shoulder and wrist than in the knee, hip, or elbow. The ankle-joint and the joints of the fingers and toes are still less sensitive. In right-handed individuals the movement sense is more delicate on the right side than on the left.

3. *Sensations of Resistance and of Weight.*—The sense of weight is not identical with that of resistance and is more delicate in the upper than in the lower extremities.

4. *Stereognostic Sense or Perception.*—The capacity to recognize the form and consistence of objects by touching and feeling them is known as stereognosis. This capacity involves the combined activities of the movement sense, resistance sense, position sense, sense of touch, and sense of temperature. Even more, certain higher cortical faculties are involved, including the spatial and temporal arrangement of the sense impressions and the memory pictures of previous sensations. Obviously, we have to do here not simply with sensation, but with a combination of sensory, associative, and reproductive processes.

5. *Vibration Sense.*—Rapidly vibrating objects when placed upon the skin over a bone yield a peculiar vibrating sensation known as pallesthesia. Clinically, one examines the sensation best by placing the handle of a vibrating tuning-fork of 64 or 128 oscillations per second firmly upon the surface of the skin over one of the bones.

**Vestibular and Allied Senses. Sense of Static Equilibrium. Sensations of Position of the Body as a Whole. Sensations of Movement of the Body as a Whole. Sensations of Orientation in Terrestrial Space. Sensibility to Accelerations in a Straight Line and to Angular Accelerations.**—Here we have to deal with a great group of centripetal impulses which are of high importance for the functions of equilibrium, locomotion, and orientation. It seems likely that most of the centripetal impulses concerned are infraconscious, or that their psychic correlate is relatively insignificant.

The directions up and down are recognized by everybody, though how the force of gravity is made known to us is not clear. Even under water the capacity for orientation as to the vertical direction is unimpaired. The mechanism, whatever it is, is sensitive to *accelerations in a straight line*, and gives rise not only to our knowledge of the vertical direction, but also to sensations of progressive or non-rotary movements. In addition to this mechanism, there is another apparatus in the head



which is sensitive to *angular accelerations* and gives us sensations of change of velocity of rotary movements. These sensations, which follow accelerations in a straight line and angular accelerations, are associated with characteristic eye-muscle reflexes. An alteration of the position of the head relative to the vertical causes a compensatory change in the setting of the eyeballs, and if the body be rotated around its vertical axis the eyes show movements of nystagmus as long as the sensations of rotation are felt.

These sensations are connected with the functions of the membranous labyrinth of the internal ear. It seems probable that the information which the brain receives regarding the position of the head when it is at rest and regarding the acceleration or retardation of progressive (non-rotary) movements depends upon the nerve endings in the utricle and saccule, upon the maculae of which the little ear-stones or otoconia rest (Breuer), while the sense organ underlying the sensations dependent upon angular accelerations appears to be situated in the ampullary crests of the membranous ampullae of the semicircular ducts. While both the utricle and saccule, on the one hand, and the semicircular ducts, on the other, appear to have to do with static as well as dynamic sensations, it seems probable that the static are mediated rather more by the former, the dynamic rather more by the latter. The loss of orientation as to the vertical when under water and the absence of vertigo on rotation so often seen in deaf mutes are thought to be due either to imperfection of the otoconic apparatus or of the semicircular ducts.

**Other Interior Senses.—Hunger.**—In its mild form this sensation is familiar to everyone as the appetite for food localized in the gastric region. The hunger sense is to some extent dependent upon metabolism. The well-known increase of appetite after muscular exertion, after exposure to cold, and in diabetes mellitus makes this clear. On fasting, the sense of hunger soon disappears, in marked contrast with the sense of thirst.

**Thirst.**—The sense of thirst is referred to the back of the throat, and if no water be taken the sensations become painful.

**Sense of Fatigue.**—If the muscles are made to contract beyond a certain point, a feeling of fatigue is encountered. Whether this is due to centripetal impulses arising in the muscles themselves or to a change in the nerve centres in the cerebral cortex is not certain.

**Sexual Sense.**—The impulse to sexual gratification is technically known as *libido*. The sexual impulse of adults appears to be a unification of various childhood impulses; these culminate normally in an effort toward a single specific goal. Originally auto-erotic, the impulses become later hetero-erotic.

**The Exterior Senses.—Touch or Pressure.**—The skin contains sensory mechanisms that permit us to recognize very slight contact and very light degrees of pressure. The sense organs are closely related to the hairs in parts of the body which possess these and to Meissner's corpuscles in other parts. When delicate stimuli are employed it is found that sensations are set up only when definite points are pressed upon. These are the so-called *touch points* of von Frey. They are much more numerous than cold points and warm points, but are less numerous than pain

points. They are very numerous on the lips, tongue, and finger tips, and are relatively much scantier on the leg and upper arm. Touch sensations following brief stimuli last only a short time, and if stimuli be repeated rapidly one receives a corresponding number of sensations without fusion in consciousness. The threshold stimulus for individual touch points varies. One can determine the threshold most easily by the use of von Frey's test hairs.

No marked feeling tone accompanies touch sensations, a fact which distinguishes them sharply from sensations of pain. The time elapsing from the moment of application of the stimulus to a touch organ to the moment of appearance of sensation in consciousness is shorter for touch than for temperature or pain (reaction time). Touch sensations can be very accurately localized upon the surface of the body; indeed, the sense of touch, together with sight and movement sense, has done most to give to man his conceptions of spatial relations. Each touch point on the surface of the body seems to have its own "local sign," that is, the sensation from it can be distinguished from that from every other. If two touch points are close together, however, they must be stimulated successively and not simultaneously if the two separate sensations are to be recognized (*successive threshold* of von Frey). When touch points are a certain distance apart, stimulation of them simultaneously gives rise to two separate sensations (*simultaneous duplicity threshold* of von Frey; *space threshold* of Weber).

**Tickling and Itching.**—These are as yet not well understood. Tickling is probably to be separated physiologically from itching. However, both are closely related to touch sense and pain sense. It is possible that they should be regarded as secondary sensations rather than primary (H. Quincke), and that they are associated with vasomotor reflexes. It is interesting that the sensations are aroused by slight stimuli and can often be inhibited by more powerful stimulation in the same areas.

**The Temperature Sense.**—In the skin and mucous membranes there are at least three varieties of sensory nerve beginnings which respond to thermal stimuli; they are connected with the so-called cold points, warm points, and pain points. The adequate stimulus for a cold point is cold, that for a warm point is warmth, but both sets of points react to various disparate stimuli also; thus one gets a sensation of cold when he stimulates a cold point by a sharpened stick, by the application of menthol, or by an electrical current, and a warm point can be stimulated not only by heat, but by mechanical irritation with fur, by exposure to carbon dioxide, or by electricity. Pain points are stimulated by cold below a certain temperature or by heat above a certain temperature. Touch points appear to be uninfluenced by thermal stimuli. The cold points are far more numerous than the warm points, but are less evenly distributed than the touch points and the pain points.

Natural stimuli, as a rule, affect areas of the skin which contain a number of sensory points, so that our ordinary cutaneous sensations are complexes made up of a number and variety of elementary sensations. When the thermal stimulus corresponds in temperature to that of the skin no sensation at all is felt; if it correspond to a temperature slightly

lower than that of the skin, the cold points alone respond and a sensation of coolness results; if it corresponds to a temperature a little above that of the skin, a sensation of warmth is felt. Very low temperatures affect both the pain points and the cold points; very high temperatures may affect the warm points, the cold points, and the pain points simultaneously, and yield a mixed sensation compounded of warmth, cold, and pain, usually designated "burning hot."

**Taste.**—There are four simple elementary taste qualities: sour, sweet, salty, and bitter. All other so-called taste sensations are either combinations of these or are mixtures of taste sensations with sensations of smell, touch, temperature, or pain. Some taste sensations are associated with pleasurable feeling tone, others with negative feeling tone. Substances which can be tasted are known as "sapid substances," and are soluble in water or in saliva. They are crystalloid in nature, colloids being devoid of taste.

Apparently there are entirely different sense organs for sour, sweet, salty, and bitter sensations. The adequate stimulus for arousing sour sensations appears to be the hydrogen ion of acids. No common chemical character has as yet been ascribed to the sapid substances which arouse sweet sensations; salty sensations are best aroused by a 2 per cent. solution of sodium chloride, though various other salts can stimulate the sense organ concerned. Bitter sensations are aroused by very different chemical substances. Many of the alkaloids (strychnine) are bitter, but numerous sapid substances are bitter which are chemically unrelated to these (*e. g.*, magnesium sulphate). The taste organs thus acted upon by chemical stimuli do not respond to mechanical or thermal stimuli. They do, however, sometimes respond to electric currents (so-called *electric taste*).

Single taste modalities are variably represented in various parts of the tongue; thus, sweet taste is best represented at the tip, salty at the tip and margins, sour at the margins, and bitter at the posterior part of the dorsum linguæ. The centripetal nerves of taste leave the tongue by way of the lingual branch of the trigeminal nerve and by way of the glossopharyngeal nerve. The few taste buds of the larynx belong to the pneumogastric nerve.

**Smell.**—This is closely related to taste sense, and naïve observers often confound sensations of smell with those of taste. The nose possesses touch sense, temperature sense, and pain sense; when we inhale formalin, it is the pain sense which is chiefly aroused, and when we inhale menthol the temperature sense is also affected. Aside from distinguishing sensations of smell proper from other modalities of sensation, we have the still greater difficulty of analyzing olfactory sensations among themselves and resolving them into their simpler psychological constituents. The olfactory sensations are mediated by the neuro-epithelial cells in the olfactory region high up in the nose.

Smell is to be regarded as a chemical sense, and the adequate stimuli are chemical substances which act directly upon the peripheral olfactory neurones. Very minute amounts of certain chemical substances are sufficient to stimulate the sense organ, and they must be gaseous or volatile in order that they may mix with the air that enters the cavity

of the nose. For human beings the smell sensations aroused by the entrance of chemical substances through the nasopharynx are more important than those dependent upon substances entering through the front of the nose.

While there are only four fundamental varieties of taste sensations, it seems likely that there are, at least, twenty or thirty elementary olfactory sensations; the difficulties surrounding experimental investigation prevent any dogmatic statement concerning this number.

Attempts at quantitative methods of examination of the sense of smell have been made, especially by Zwaardemaker. The term *olfactometry* has been applied to the measurement of the sensibility of the olfactory organ for adequate stimuli, and the term *odorimetry* is used to indicate the making of comparative measurements of the stimulating action of different substances.

We know nothing of exact localization of olfactory sensations. The olfactory sense is not easily fatigued, although it varies in this regard in its different modalities. The close relation of the olfactory sense to various reflex, instinctive, and affective phenomena need only be mentioned to be appreciated. The odor of food excites the impulse to eat and starts the secretion of gastric juice. The relation of the sense of smell to the excitation of the sexual impulses is well known, and the relation of odors to various affective states is close. The mood may be changed in a moment by a scent or a stink.

The clinical examination of the sense of smell is usually carried out by holding odoriferous substances before one nostril at a time; the patient inhales and tries to name the substance. Well-known substances, chiefly, are used. As the majority of people are surprisingly inaccurate in the discrimination of smell sensations, the following series can be recommended: Rubber, gutta-percha, ether, yellow beeswax, camphor, oil of anise, vanilla, musk, asafetida, cacodyl or ammonium sulphide, roasted coffee, pyridin, tar or carbolic acid, cheese or mutton tallow, tincture of valerian, skatol.

Where smell is defective a careful examination of the nose should be made to make sure that the defect is not due to obstruction of the air currents in the nose rather than to any disturbance of the nervous mechanism itself. In addition to trying the olfactory sense, one may, if he likes, test the functions of the trigeminus in the nasal mucosa with the fumes of acetic acid, ammonia, formalin, and menthol.

**Hearing.**—The organ of hearing mediates the sensations of sound. Sounds are divided into two great groups known as *musical sounds* and *noises*. The vibrations reach the organ of Corti either by *air conduction* through the external and middle ear or by *osteotympanal conduction*, the transmission occurring then through the bones of the skull. The organ of hearing is attuned to certain periodic movements only; thus, oscillations less frequent than 16 per second or more frequent than 50,000 per second fail to stimulate the ear.

Sounds are ordinarily heard with both ears (*diotic hearing*), though sometimes with one alone (*monotic hearing*). If a tuning-fork (*c-c'*) be placed on the mastoid, the sound is heard by osteotympanal conduction. If at the moment when the sound ceases the fork be removed from the

mastoid and placed before the ear, the tone can again be heard through air conduction (*Rinne's experiment*). If a vibrating fork be placed in the median line of the head, exciting the two ears equally, and then one ear be loosely closed with the palm of the hand, the sound is markedly intensified in the closed ear—so-called lateralization of the sound (*Weber's experiment*). In otiatric diagnosis it is often important to determine the *upper and lower limits of audible tones*. The best apparatus for this is the "continuous tone series" of Bezold-Edelmann, which permits of stimulating with vibrations numbering from 16 to 48,000 per second.

*Clinical Examination of the Sense of Hearing.*—Ordinarily clinicians are satisfied with (1) determining the so-called auditory acuity; (2) testing the upper and lower tone limits; (3) Rinne's test; (4) Schwabach's test; (5) Weber's test; (6) Gelle's test.

To determine the *auditory acuity*, various rough-and-ready tests, serving for general orientation as to hearing power, have been introduced. One tests the ear first for sounds of different kinds. Thus the power to hear *spoken* and especially *whispered* words is examined, preferably in a still, closed room. The average distance at which whispered sounds can be heard (the whisper being produced with the aid of the residual air left in the thorax after deep expiration) is about 25 meters in a very quiet room, about 20 meters in ordinary rooms. A patient whose hearing permits these words to be heard as far away as 6 meters suffers no practical inconvenience. Each ear is tested separately, the patient standing at the end of the room, the other ear being tightly closed with the finger and the face and eyes looking straight forward so as to render lip reading impossible. Beginning at a distance of 6 meters a word is whispered, and the distance gradually decreased until the word is recognized and correctly repeated.

If whispered words are not heard at all, one uses "conversational tones" in the same way. Spoken words can, however, be heard, when one ear is deaf, at a distance of 6 meters, even when the healthy ear is closed. To be sure about this, if the patient hears conversational tones at a distance of 3 meters when the good ear is closed and the bad ear open, one tries again when he closes also his bad ear (*Lucae-Dennert test*); if he can no longer repeat the test words, this proves that he heard with the bad ear, but if he repeats them one can be sure that the bad ear did not hear. We can now exclude hearing in one ear entirely by the use of Bárány's "noise-apparatus."

More accurate are measurements with Politzer's *acumeter*, an instrument which yields sounds of equal intensity; the drawback to it is that the noise it produces consists exclusively of a few high tones. For the same purpose the tick of a watch may be used. But the distance at which sound can be heard is not a good measure of auditory acuity.

The poorer the hearing in one ear, the more quickly a waning tone ceases to be perceived, and the measurement of the time a tone is heard is, therefore, an important aid in otiatric diagnosis. The numerical expression of defective auditory acuity is not simple. If a patient, for example, hears the fork *c* for 92 seconds, while a normal ear hears it 183 seconds, the auditory acuity under test (*H P*) is not one-half the normal acuity (*H N*) but a much smaller fraction, about  $\frac{1}{3\frac{1}{2}}$  (Ostmann). This depends on

the fact that during the waning of vibrations of a fork the intensity does not diminish by equal amounts in successive seconds; at first the intensity decreases rapidly, then ever more slowly.

But for any measurement of auditory acuity which pretends to accuracy one must be able easily (1) to use tones of any desired pitch, (2) to use pure tones, (3) to measure their intensity exactly, and (4) to offer them to the ear in even intensity for any desired period. The instrumentarium which best meets these requirements is that of Wien, in which a telephone is used as a source for the tones, the sinusoidal alternating currents being yielded by a sinus inductor (for low tones) and by an alternating current siren (for higher tones).

Having determined the *degree* of auditory defect, one has next to try to ascertain its *cause*; one must decide whether the defect is due to the *sound-conducting apparatus* (auditory meatus, tympanic membrane, middle ear) or to the *sound-perceiving apparatus* in the internal ear. The data to be gathered consist in the following:

1. *Determination of upper and lower tone limits* by means of the Bezold-Edelmann continuous tone series or more simply by tuning-forks  $c_1, c, c_4$ ; the upper limit may be normal and the lower limit changed in middle-ear disease, while the opposite holds for labyrinthine deafness.

2. *Rinne's Test*.—The difference between the time of air conduction and of bone conduction should be noted in seconds for a fork having a pitch somewhere between  $c$  and  $c^2$ .

3. *Schwabach's Test*.—Comparison of the time a fork is perceived when held on the vertex with the time in a person of normal hearing; the time is shortened in labyrinthine disease.

4. *Weber's Test*.—The sound may be "lateralized" in one ear without artificial closure—toward the deaf ear in middle-ear disease, toward the healthy ear in labyrinthine deafness.

5. *Gelle's Test*.—If one compresses the air in one meatus by means of a rubber bulb, the tone of a fork held on the same half of the skull will be less loud at the moment of compression if the base of the stirrup be movable, while it will remain unchanged if it be fixed (ankylosis).

**Sight.**—When the eyes are open the visual sensations are spatially extended and exteriorized as the so-called *binocular visual field*. If one eye is closed we project a *monocular field*. When both eyes are closed, or if they be open in the dark, there is a sense of blackness, something quite different from the absence of sensation corresponding to the region peripheral to the visual fields.

There are two series of visual sensations: (1) An achromatic series—black, gray, white—which can be represented geometrically in their transitions by a straight line; and (2) a chromatic series—red, orange, yellow, green, blue, violet, purple (in all at least 160 tones at full saturation)—which can be represented geometrically in their transitions by a line which returns to the starting point, *e. g.*, by a circle. This series of sensation is related to the achromatic series in such a way that the greatest saturation of each color occurs normally at a certain luminosity, *e. g.*, yellow is brighter than orange when each is fully saturated. This relation of the chromatic to the achromatic series gives rise to the so-called

variables in sensations of color. There are three conditions of variability—the first dependent on quality, the second on luminosity, the third on saturation.

The organ of visual sense includes the eye and a series of neurone chains in the central nervous system. The *dioptric* apparatus of the eye determines the mode of application of the *adequate stimuli* (light rays) to the sense organ proper, the *retina*. The impulses arise in the rods and cones and pass through the bipolar cells and the ganglion cells of the retina into the optic nerve and thence to the primary and secondary optic centres of the brain. The whole path from the retina to the cortex, composed of several sets of superimposed centripetal neurones, is known as the *visual conduction path*.

It is interesting that the different portions of the retina behave very differently in regard to color sensations; thus, at the fovea (*central vision*) and the area immediately adjacent (*paracentral vision*) the color sense is quite different from that of the more peripheral portions of the retina (*excentric vision*). Objects which seem colored in central vision may have different colors in paracentral vision, and may be colorless when excentrically viewed. In other words, the periphery of the retina is totally color blind and its vision is monochromatic; the central portion of the retina has polychromatic vision, while there is an intermediate zone which is partially color blind, having dichromatic vision. This explains why it is that the visual fields for colors are of different sizes even in the normal eye.

The ether vibrations of the light rays lead to photochemical, histological, and electrical changes in the retina and give rise to the nervous impulses. The luminosity of a visual sensation is a function of the actual energy of the ether vibrations and of the parts of the retina stimulated. The eye cannot analyze white into its constituent components, and differs from the ear, which can recognize the partial tones in a clang.

*Color blindness* is a deviation from the normal relations in a system of color sensations. It may be most properly defined simply as abnormal color sense. In its least abnormal form it manifests itself in a general weakening of the color sense, while in its most abnormal form no color can be seen at all but only differences in luminosity are distinguished. Between these extremes various deviations from the normal occur.

The following may serve as a classification of such abnormal cases.<sup>1</sup>

1. *Abnormal polychromates*, who see many colors but who differ from normal individuals (a) in that one or more color qualities are absent or (b) in outstanding color relations (*e. g.*, in the similarity of certain colors to one another). These are the most common cases, and in the majority it is the colors red and green which are not distinguished.

2. *Dichromates*, who see only one pair of colors, these being complementary, though sometimes different from the normal complementary colors.

3. *Achromates*, who see no colors at all, all lights giving sensations of gray in varying intensity. These are extremely rare.

<sup>1</sup> A. Kirschmann, *Arch. f. d. gesamte Psychol.*, 1907

Visual sensations depend not only upon the stimuli applied to the eye, but also upon the state of the eye at the time of application of the stimulus. The change in the eye in the different phases of activity is known as its power of adaptation. Among the phenomena which pertain to this function of adaptation are: (1) Positive and negative images; (2) light induction and color contrasts; and (3) the adaptations undergone by the eye in the light and in the dark. Studies of the *dark-adapted eye* make it probable that the rods of the retina are especially concerned with vision in dim light (night), while the cones of the retina have to do rather with vision in strong light (day).

*Localization of Visual Stimuli.*—Two retinal stimuli which have the same value as regards quality and luminosity may differ in that they are projected into different parts of the visual field. All the retinal excitations taken together give rise in consciousness to the continuous extended field. We concentrate our attention normally upon the point upon which our eyes focus; while the more peripheral portions of our visual fields are less focal in consciousness. The arrangement in space of visual perceptions, therefore, depends upon the local sign which adheres to the sensation aroused by stimulation of each retinal point. The localization depends, however, not only upon this local sign, but especially as concerns measurement, also upon eye-muscle sensations.

In examining a patient clinically regarding his capacity for visual sensations we are provided with very exact methods and delicate apparatus. Here we can call attention only to some of the methods most essential for neurological diagnosis, especially to (1) the determination of visual acuity, (2) the determination of the type of central vision, (3) the determination of the capacity for vision in dim lights, (4) the determination of the extent of the visual fields, and (5) the ophthalmoscopic examination.

In *determining the visual acuity*, one uses letters or similar signs, made so as to be as long as they are broad; the strokes of the letters should be of even thickness and one-fifth of the thickness of the whole letter (*e. g.*, Snellen's "Optotypi"). The visual acuity  $V$  (*visus*) is expressed by a fraction whose numerator gives the distance  $d$  in which the letter is recognized, and whose denominator gives the distance  $D$  which corresponds to a visual angle of five minutes for the letter:  $V = \frac{d}{D}$ . The dioptric apparatus must be borne in mind (anomalies of refraction, accommodation), and if faulty, the visual acuity determined before and after correction.

In *determining the types of central vision* we decide whether the patient has polychromatic (normal), dichromatic, or achromatic vision. For this Holmgren's colored wools are very satisfactory. From a large collection of wools of different color one chooses one of rose-color (pale bluish red) and asks the patient to put beside it from the collection all the wools of the same color, telling him that he may make mistakes either by choosing wools of a wrong color or leaving behind wools of the same color. If his vision be dichromatic, the patient will pick out bluish green and gray wools and put them with the rose-colored wools. If his vision be achromatic, he will be wholly unable to discriminate among the wools



except by differences in brightness. One may use Adler's colored crayons or Stilling's pseudo-isochromatic charts, Nagel's lantern with colored lights, or color equations with rotating disks.

In *determining the capacity for vision in dim lights*, one darkens the room and finds out whether the patient sees as long as the examiner and whether his eye becomes dark-adapted as quickly. For quantitative tests, Foerster's photometer or Masson's disk may be employed.

The *determination of the extent of the visual field* for each eye (perimetry) is of very great importance in neurological diagnosis. A *rough test* may be made by seating the patient with his back near a window before the examiner; a bandage is placed over one eye and he is told to look with the other into the eye of the examiner opposite (distance, 40 cm.). Closing one eye, one holds the white or colored object (1 cm.) midway between the patient and the examiner and controls by the extent of his own visual field the examination of that of the patient. With a little practice one can detect existing hemianopsias, scotomata, or concentric contractions easily; the method is important for quick preliminary orientation.

For exact registration of the extent of the visual fields, Foerster's *perimeter* or some similar apparatus (*e. g.*, Sydow's) should be used; self-registering perimeters are obtainable, but are not necessary. Care must be taken to avoid fatigue; one should not work too long at one visual field, but determine limits quickly (though accurately), encouraging the patient frequently to give the matter his closest attention. The patient is told that while he may wink as often as he likes, he is to look straight forward at a fixed point and on no account to follow the object with his eye. The objects are brought in from the periphery and the patient announces the time of first appearance and the color; white objects are brought in vibrating, colored objects quietly, from the periphery. No one color field should be determined all at once, but one tries different colors, so that the patient never knows which color is entering the visual field. If a defect (scotoma) be found, its limits may be very accurately localized by using small test objects (2 mm. broad). For testing *color fields*, one usually uses green, red, and blue, the field for the former being smallest, that for the latter largest, under normal conditions.

There are great individual differences, however, in normal color fields, and much depends upon the exact colors used. If a green which is complementary for a red be employed, the fields for red and green are approximately co-extensive.

The technique of *ophthalmoscopic examination* should be learned and regularly practised by every active physician. Through it we are able to look directly at the papilla N. optici, a part of the central nervous system, and, from it, to draw conclusions regarding a whole series of nervous diseases; it is one of the chief aids to neurological diagnosis. The examination in the *inverted image* is more desirable than in the *upright image*, since the technique is easier and a wider view of the retina is afforded; the upright image is, however, very valuable for the study of finer details. As to the papilla N. optici itself, one notes any changes (*a*) in the normally distinct boundaries, and (*b*) in its normally delicate rose color. It is to be borne in mind that the temporal half of the papilla

is normally of a somewhat lighter rose color than the nasal, but it is never really white. The centre of the papilla often looks white normally, owing to the "physiological" excavatio papillæ; the periphery is, however, always rose color. Acquaintance with the normal appearances of the arterioles and venules of the retina, the color of the eye-ground, and the macula lutea should be cultivated.

## DISTURBANCES OF SENSATION.

**General Remarks.**—It is customary to divide sensory disturbances into subjective and objective. By *subjective* disturbances one means those which are announced spontaneously, in the consciousness of the patient; by *objective* disturbances, those abnormalities of sensation recognized by the patient when he himself or his physician applies adequate stimuli to his organs of sense.

Subjective disturbances are nearly always pathological sensations which arise, owing to (1) abnormal internal stimuli (acting on the sense organ, conduction path, or cortex cerebri), or (2) abnormal increase in excitability of these parts. The pathological sensations may not differ in quality from the sensations producible by adequate stimuli; when they do differ (*e. g.*, formication) they are spoken of as *dysesthesias* or *paresthesias*.<sup>1</sup>

Objective disturbances are usually classed under the headings: (1) *Anesthesia*, (2) *hyperesthesia*, and (3) *paresthesia*, corresponding to (1) decrease or loss, (2) increase, and (3) perversion of sensibility. This is a very crude classification, and must sooner or later be modified and extended. A more rational classification is needed, and we shall doubtless from now on begin to classify anomalies of sensation according to changes in (1) *quality*, (2) *intensity*, (3) *reaction time*, (4) *duration*, (5) *feeling tone*, (6) *localization* or *exteriorization*, (7) *fusion*, (8) *adaptation*, (9) *fatigue*, and (10) *relation to reflex, affective, and associative processes*.

By *anesthesia* is meant a condition in which an adequate stimulus no longer arouses a sensation. It may be temporary (*e. g.*, hysteria, cocaineization) or permanent (*e. g.*, optic atrophy). It may be peripheral in its origin (sense organ, centripetal nerve) or central (conduction path, cortical sense area). Aside from the complete loss of sensation which anesthesia denotes, conditions of *hypesthesia* are met with in which the sensations aroused by adequate stimuli are less intense than normal, or the physical stimulus necessary to elicit a just perceptible sensation must be stronger than normal (elevation of intensity threshold).

In *hyperesthesia* stimuli when applied call forth sensations of greater intensity than normal or a stimulus normally subliminal possesses a threshold value (*depression of intensity threshold*). Much of what is designated hyperesthesia clinically, is really an affective state due to *abnormal negative feeling tone* (*e. g.*, the "acoustic hyperesthesia" of psychoneurotics). The "positive or pleasurable feeling tone" of other

<sup>1</sup> The term *paresthesia* is used very loosely; some mean by it abnormal sensations like formication; others use it for anomalies in reaction time, fusion, localization, etc.

sensations may be lost (*anhedonia*), diminished (*hyphedonia*), or increased (*hyperhedonia*); changes here are met with especially in mental diseases.

The *area of sensory surface* in which the anomaly of sensation exists, or so-called *distribution* or *extent* of the anesthesia or hyperesthesia, should be accurately determined and its *limits* exactly recorded, since these are of much importance in differential diagnosis, permitting one frequently to locate the lesion in periphery, spinal cord, or brain.

When an organ of sense is diseased it is important also to make sure whether the anomaly concerns all the sense modalities or sense qualities mediated by it (*total anomaly*) or only certain of them (*elective anomaly*). Thus, as an example of an elective anomaly of cutaneous sense, the condition in syringomyelia may be mentioned in which touch sense may be intact when the sense of temperature and pain is abolished.

The *apperception time* for sensation may be pathologically delayed (*e. g.*, delayed pain and touch sensations in hypesthetic, or more often hyperesthetic, areas in neuritis and tabes).

The *localization* and *exteriorization* of sensations are sometimes abnormal. Irritation of a nerve trunk is, according to the "law of excentric localization," always referred to the region whence the nerve receives normally its centripetal impulses; thus, when one "hits his funny-bone," the little finger tingles, and in amputation neuromata the disagreeable sensations are referred to the lost member. Again, a stimulus applied to one part of a sensory surface may be localized in another; in tabes, hysteria, and multiple sclerosis a skin stimulus to one leg may be localized in the other (*alloecheiria* or *allescnesia*). When a stimulus applied to one point is properly localized there but gives rise also to a subjective sensation (perhaps of wholly different modality) localized elsewhere we speak of *synesthesia*.<sup>1</sup> There may be anomalies of *fusion* of sensations; stimuli which should be felt as continuous and simultaneous may be experienced as successive, and one touch to the skin felt as two or more, or two compass-points as three or four (*polyesthesia*).

**Disturbances in Individual Sense Domains.—Anomalies of Pain Sense.**—Subjective anomalies are very common. Spontaneous pains occur in various organic diseases (*e. g.*, neuritis, tabes, visceral diseases), as well as in so-called functional diseases (psychoneuroses). Neuralgias, cephalalgias, hemicranias, visceralgias, and topoalgias or psychalgias are symptoms met with daily. In studying abnormal pain sensations, the elements of the pain should be carefully analyzed, its relations to space and to time, the exact quality of the pain, its topography, and any accompanying phenomena should be attended to. Violent pains on the side of the body which is the site of a hemianesthesia point to a lesion of the optic thalamus ("thalamic syndrome" of Roussy). The modifying influence of position, of motion, of pressure, of food, of drugs and chemicals, and of the organic functions of the body should be looked into. The excellent treatise of Schmidt<sup>2</sup> can be heartily recommended as a valuable guide to the interpretation of pains complained of by patients.

<sup>1</sup> Smith, H. L., *Johns Hopkins Hosp. Bull.*, 1905, xvi, 256. Here belongs the so-called sound feeling, color hearing, or sound seeing, and number form.

<sup>2</sup> *Pain; Its Causation and Diagnostic Significance in Internal Disease*. English translation by Vogel and Zinsser, 1908.

Anesthesia in the domain of the sense of pain is called *analgesia*, while hyperesthesia here is called *hyperalgesia*. When spontaneous pains appear in an anesthetic area, we speak of *analgesia dolorosa*.

**Anomalies of the Deep or Mesoblastic Sensibility.**—Anesthesia in this domain has been designated *bathyanesthesia* (Oppenheim). (1) *Anomalies of the sense of position*. Hypesthesia and anesthesia are common in neuritis and tabes. If a limb be placed in a certain position, this can be only imperfectly or not at all reproduced by the opposite limb. (2) *Anomalies of the sense of movement*. If anesthesia exists, passive movements are not felt; if hypesthesia, greater excursions than normal have to be made to arouse the sensation of movement (elevation of threshold). Not only the extent, but the direction of the movement may be mistaken in these *kinanesthesias*. As a result, there is disturbance of the coördination of movements, or ataxia. Localized loss is often cortical in origin and helps in topical diagnosis (cerebral tumor, etc.). (3) *Anomalies of the sense of resistance and weight*. This sense is prone to suffer also when the sense of movement is disturbed. Weights are improperly judged and the difference sensibility is altered. (4) *Anomalies of stereognostic perception*. In disease the stereognostic perception may be more or less impaired, even to complete loss (*astereognosis* of C. K. Mills). It may be lost even when the sense of touch, temperature, and pain are normal. Astereognosis is common in hemianesthesia of cerebral origin, but it is difficult to be sure about it unless the accompanying hemiplegia is so slight as to permit the patient to make the movements necessary in palpation. It is more likely to be present if the hemianesthesia be cortical (Regio centroparietalis) in origin rather than capsular, pontine, or bulbar. In the cerebral palsies of children the power of stereognostic perception may never develop. Astereognosis will appear in tabes and peripheral neuritis if the sensory conduction paths from the upper extremities be involved. Astereogenesis is sometimes a symptom in hysteria. (5) *Anomalies of the vibration sense (osseous sensibility)*. Anesthesia of the bones to the vibrating tuning-fork (*pallanesthesia*) is common in neuritis, tabes, syringomyelia, hematomyelia, Brown-Séquard paralysis, myelitis, cerebral hemianesthesias, and hysteria (Dejerine).

**Anomalies of the Vestibular and Allied Senses.**—**Vertiginous Disorientation.**—Disturbances of the vestibular or labyrinthine sense in its two components: (a) Sensibility to acceleration in a straight line (sense of the vertical direction and of progressive, non-rotary movements), and (b) sensibility to angular accelerations (sense of change of velocity of rotary movements)—have only recently had attention more carefully directed to them.

A remarkable phenomenon associated with disturbance (even temporary) of the vestibular sense is an *illusion regarding the state of movement of the body*, a deception as to its momentary stability, in which one has the mistaken sensation either (1) that objects in the environment are revolving around one's body, or (2) that the body is rotating around one of its axes. This sensation, often accompanied by a decided feeling of discomfort, occasionally, even with the nausea or vomiting, constitutes the form of dizziness or *vertigo* known as *sham-movement vertigo*.

(to distinguish it from height vertigo, felt on looking downward from a great height or upward on a steep wall). This sham-movement vertigo appears to be the result of a "*contradiction*" of one set of sensations by another. If one of our sense organs signals "the body is at rest," and another signals "the body is in motion," or if the signals conflict regarding movement in the environment, we feel confused, disoriented, dizzy. This *vertiginous disorientation* is most often due to a *discrepancy between the reports* given us by our vestibular sense and those emanating from other sense organs; it is usually associated with a tendency to move the body in a direction opposite to a movement really occurring or to a sham movement, and, indeed, the body is often so moved.

This feeling of vertigo can be aroused by stimulating the N. vestibuli in various ways: (1) By rotating the body around any one of its axes; (2) by moving the body progressively in a curved path (swinging; pitching of a ship); (3) by passing a galvanic current through the head from one ear to the other (galvanic vertigo). Here the abnormal vestibular sensations contradict visual, muscle-joint, and tactile sensibility. But the same sensation can be aroused when vestibular sense is normal through contradictory abnormal sensations of sight or of eye-muscle movement, *e. g.*, whenever binocular vision is suddenly disturbed (eye-muscle paralysis), or whenever the image in one eye is sharply focussed and the other is blurred. Further, in cerebral lesions (local irritation of abscess), cerebral anemia or cerebral intoxications (acute alcoholism), in all probability, the central stimuli affect sensory neurones of certain modalities more than others, so as to produce contradictory impressions with disorientation.

Recently, attempts have been made to analyze the different forms of vertigo and other disturbances in vestibular domains by means of Bárány's tests,<sup>1</sup> especially by the so-called "caloric nystagmus tests," made by injecting cold water and warm water into the external auditory canal. Cold water normally produces a nystagmus directed toward the opposite side, warm water a nystagmus toward the same side. When the vestibular paths are interrupted these normal reactions do not occur, or are altered.

Obviously, then, any factors which disturb the functions of the vestibular nerve or its central connections or the eye-muscle apparatus (peripherally or centrally) may cause vertigo, especially if the disturbance take place quickly. Many deaf mutes experience no vertigo on rotation, probably owing to loss of function of the labyrinth.

**Anomalies of Other Interior Sensations.**—*Hunger.*—Hypesthesia and anesthesia in this domain are met with in the various grades of *anorexia*; hyperesthesia, expressed as exceedingly strong feeling of hunger, is called *bulimia*, or "wolf hunger," when this is accompanied by powerfully unpleasant feeling tone, the patient describing it as painful, it is called *gastralgokenosis* (Boas).

The *sense of satiety* may be diminished or abolished (*akoria*, *aplesia*); patients (tumor cerebri, dementia) may then ingest extraordinary amounts of food and yet remain entirely unsatisfied; they exhibit

<sup>1</sup> Lewandowsky, *Handb. d. Neurol.*, Berlin, 1910.

*polyphagia*. The feeling of *nausea* is probably an affective state rather than a sensory disturbance in the strict sense. *Gastralgia* (including *cardialgia* and *pyrosis*) is a disturbance of visceral pain sense.

*Thirst*.—Abnormal thirst occurs in diabetes and often also in the psychoneuroses; it may lead to excessive drinking (*polydipsia*), and is probably central in origin.

*Fatigue*.—Under abnormal feelings of fatigue are often grouped three distinct varieties of sensations: (1) The *painful weariness* or *feeling of being completely "knocked out,"* which is associated even with slight exercise in many nervous patients; (2) the *weakness* which is experienced by patients who have paralysis or paresis of muscles; and (3) the *general lassitude* felt on slight exercise in states of exhaustion, in which slight movements cause palpitation, dyspnoea, perspiration, tremor, and faintness.

*Sexual Sense*.—Anesthesia of the sexual sense (abnormal *frigidity*) may be present owing either to *loss of libido* or to *dyspareunia*; the libido may be pathologically increased, as in *nymphomania* and *satyriasis*; for various *perversions of the libidos*, special texts may be consulted. The hyperesthesia of the vagina (with painful contraction of the constrictor cunni), known as *vaginismus*, is an anomaly of visceral pain sense and nearly always associated with abnormal affective states.

**Anomalies of the Sense of Touch or Pressure.**—Anesthesia and hypesthesia of the touch sense very frequently occur; it is doubtful whether true hyperesthesia of this sense has been observed. The distribution of the anesthesia or hypesthesia is important for diagnosis, as is association with (or dissociation from) anomalies of pain and temperature sense.

Spontaneous itching (*pruritus*) is regarded as a dysesthesia. Special forms of formication are met with in *acroparesthesia* (tingling and pain at ends of extremities), in *meralgia paresthetica* (dysesthesia in the domain of N. femoris lateralis), and in *erythromelalgia*.

**Anomalies of the Temperature Senses.**—Subjective feelings of chilliness or cold or heat are common in infections (*e. g.*, initial chill), and in the psychoneuroses ("hot flushes"). The feeling of heat (especially in the back) which occurs in paralysis agitans is well known, and patients suffering from exophthalmic goitre frequently complain of abnormal sensations of heat. In *cutis anserina* the "chilliness" contains besides its cold component also a factor due to the stimulation of the touch points by contraction of the muscoli pilorum. In objective disturbances the *thermanesthesias* usually involve cold sense and warmth sense simultaneously, but this is not necessarily the case; there may be loss of cold sense (*kryanesthesia* or *alganesthesia*) without change in warmth sense, or there may be loss of warmth sense (*ardanesthesia*) without involvement of cold sense, the latter dissociation being more frequent than the former. *Thermhypesthesias* are common; *thermhyperesthesia* is rare, much that is described as such being in reality hyperalgesia.

**Topographical Distribution of Disturbances of Sensation.**—Disturbances of cutaneous and deep sensibility, especially studied accurately as to their topographical distribution on the surface of the body, throw much light upon the site of the lesions which give rise to them. In the peripheral nerve trunks and in the roots of the spinal and cerebral nerves

the fibres which carry centripetal impulses are more intimately mixed with one another than are their intracentral communications, and this explains why it is that dissociations, although they occur in peripheral and radicular lesions, are more common when the pathological process is located in the spinal cord or cerebrum.

**Anomalies of the Sense of Taste.**—Anesthesia in this domain is known as *ageusia*, hypesthesia as *hypogeusia*, and hyperesthesia as *hypergeusia*. If one or more of the four fundamental qualities of taste sensations remain intact when others are abolished, we speak of an *elective ageusia*. Various perversions have been described under the designation *parageusia*.

**Anomalies of the Sense of Smell.**—Here, similarly, we have to deal with *anosmia*, *hyposmia*, and *hyperosmia*, which may be total or elective. When an anosmia is due to interference with the passage of odoriferous air currents toward the regio olfactoria it is called an *anosmia respiratoria*, a subdivision of which is the *anosmia gustatoria* when the odoriferous currents from food and drink by way of the choanæ are obstructed. Anosmia may be due also to disease of the regio olfactoria itself (tobacco poisoning; syphilis) or to intracranial lesion of the olfactory nerves, bulbs, tracts, or centres. The left-sided anosmia which sometimes accompanies right-sided hemiplegia and aphasia is said to be due to lesion of the lateral root of the olfactory tract. In hysteria a unilateral “functional” anosmia frequently accompanies, or rather forms a part of, the hysterical hemianesthesia. Anosmia may be a congenital anomaly, and hyposmia is common in imbeciles, although not constant, some imbeciles appearing to be hyperosmic.

**Anomalies of the Sense of Hearing.**—Total deafness is known as *anacusis*, partial as *hypacusis*, while abnormally sharp auditory acuity, rarely met with, is *hyperacusis* or *true hyperesthesia acustica*.<sup>1</sup> The painful sensations in the ear felt when noises or tones are heard not only in diseases of the middle and internal ear, in otitic paralysis of the N. facialis, in trigeminal neuralgia, in migraine, and in many psychoneurotic states has been called *dysacusis* (*false hyperesthesia acustica*); in facial paralysis it takes especially the form of increased sensitiveness to noises, and has been called *oxyakoa*.

But little is known as yet regarding auditory disturbances due to *lesions of the central auditory paths*; the latter, owing to the semidecussations at the corpus trapezoideum, are laid down bilaterally, the lemniscus lateralis, etc., on each side containing conduction paths from both ears. Focal lesions in the pons and midbrain (involving the lemniscus lateralis or the collicula inferior) or higher up (involving the medial geniculate bodies or the auditory radiations) cause disturbances of hearing on both sides (not complete deafness on either side). Total unilateral deafness is sometimes caused by *neurofibromata of the N. acusticus*, but usually only after the tumor has attained to a considerable size; the deafness is here due chiefly to the lesion of the peripheral (not the central) auditory neurones. Cortical lesions involving the auditory sense areas in both temporal lobes must be widespread to cause total “*cortical deafness*.”

<sup>1</sup> The latter term has unfortunately been applied to the pain, or negative feeling-tone, experienced by psychoneurotics when noises or any sounds are heard.

**Anomalies of the Sense of Sight.**—Anesthesia in this domain is known as *anopsia*, hypesthesia as *hypopsia*, and hyperesthesia as *hyperopsia*.

Anopsia (*amaurosis* or *blindness*) or *hypopsia* (*amblyopia*) may involve the whole of one eye or of both eyes. Or one-half of each retina or its conduction paths may be blind, leading to loss of half of the visual fields (*hemianopsia*); if it be the two right halves or the two left halves of the visual field which are lost, we speak of *hemianopsia homonyma* (due to lesions of visual sense area in the cortex, of occipitothalamic radiation, or of one tractus opticus), while if it be the two medial (nasal) halves or the two lateral (temporal) halves (the latter due to chiasm lesion) of the field, we call it *hemianopsia heteronyma*. If a hemianopsia involve the color sense only or chiefly, it is called a *hemichromatopsia*; if vision be present in half the field but dimmed, the condition is designated *hemiambyopia*. A hemianopsia is said to be *total* when the line separating the blind area from the area in which the field is still visible passes perpendicularly through the fixation point; if the visual field is not so limited, but encroaches upon the blind area, the hemianopsia is *not total* and the area of encroachment is called the *surplus field* (Wilbrand). Sometimes this surplus field is so large as to occupy a whole quadrant of the visual field and then a *quadrantic hemianopsia* is spoken of. There are even references in the bibliography to sextantic and octantic hemianopsis; when the loss in the visual field is as small as this, however, it is probably better to call it a *scotoma*, as this name is applied to small areas of loss or defect in the visual field, such as the *blind spot*, which exists normally. It is customary, however, to reserve the term *scotoma* for a defect in the visual field which does not reach the periphery. A *central scotoma* is one situated at or near the fixation point; if it involves only sensations of red and green, it is called a *relative scotoma* (e. g., in chronic form of retrobulbar neuritis, limited to the papillomacular bundle, most often seen in alcoholism or tobacco poisoning), while if no light be sensed in the defect it is said to be an *absolute scotoma* (met with in diabetes, multiple sclerosis, and occasionally "idiopathically").

When the visual field is restricted regularly at the periphery we speak of *concentric restriction or contraction of the visual fields*; when this is due to progressive diseases of the Nn. optici (primary and secondary optic atrophy) the contraction proceeds from the periphery centralward, the central acuity for white and colors (especially green and red) also suffering early; the vision is "fogged;" then marked disturbances of orientation supervene. The beginning and progress may be very insidious in simple (so-called primary) progressive atrophy (tabes; dementia paralytica), while the foggiess and darkness may come suddenly in neuritis (meningitis; lues). In choked disk (tumor, lues, brain abscess, hydrocephalus) vision may remain good for some time; if the cause persists, there will be gradual loss, due to atrophy; the color sense may go first, and, finally, there may be complete loss of the visual field.

It is interesting that the contraction of a visual field due to glaucoma is especially marked in the medial field, the last of the field to go being an excentric temporal area (*glaucomatous visual-field slit*); in contrast to what happens in simple optic nerve atrophy the color sense remains



long intact in the retained field and the central visual acuity is longer maintained (Axenfeld).

In detached retina there is contraction of the field corresponding to the degree of detachment; this may in daylight affect, at first, colors only, especially blue, although in dim light the contraction is absolute.

Recent retinitis due to nephritis or diabetes frequently injures central vision and may give rise to central scotomata of different kinds corresponding to the various patches and hemorrhages; the periphery is less often injured.

In retinitis pigmentosa and in the retinal atrophy following syphilitic chorioretinitis there may be either marked concentric contraction of the visual field, or, in cases in which the degeneration attacks an intermediary zone of the retina, a so-called *ring scotoma* which later extends both peripheralward and centralward. Central vision may, despite this, be long retained as an "island" of good visual acuity and good color vision, the patient seeing as though through a tube of small caliber. Similar limitations of the visual field are met with in hysteria and in traumatic neurosis; here, however, by testing first at a distance of 10 cm. (and marking limits with chalk) and then at a distance of a meter, the visual field does not correspondingly enlarge, the psychoneurotic state here behaving like that of a stimulant.

In contrast with the *immobile positive scotomata* (due to diseases of the retina or visual path) above described, the *mobile positive scotomata* (movable shadows) due to turbidity of the corpus vitreus should be borne in mind. In neurasthenics and myopics these *muscæ volitantes* may be present when the vitreus is normal (*myodesopsia*); the threads, flocculi, veils, chains, etc., seen in certain cases may, however, be due to pathologically formed elements in the vitreous visible with the ophthalmoscope. Important for neurologists, too, is the so-called *fortification scotoma* of migraine; a positive and sharply localized sensation of glimmering followed by darkening due to irritation of the retina or visual sense area.

Sudden *blindness of one eye* may be due to embolism or thrombosis of the *arteria centralis retinae*. Other *sudden disturbances of vision* may occur in (1) cerebral hemorrhage involving the central visual paths (hemianopsia associated with other symptoms of cerebral apoplexy); (2) retinal detachment (disturbance beginning at periphery of visual field, preceded by characteristic irritative phenomena); (3) choked disk (obscurations temporary). Less sudden but rapid blindness may occur in (1) very acute inflammatory glaucoma (excruciating unilateral headache); (2) large hemorrhages into the corpus vitreum (red or green stripes before the eye precede the blindness); and (3) acute retrobulbar neuritis.

The terms *nyctalopia* and *hemeralopia* need explanation. When a patient can see better in dim light (or when the eye is in shadow) than in bright light he has *nyctalopia* (e. g., in beginning cataract, the widening of the pupil in dim light leaving more transparent lens uncovered). When, on the other hand, a patient does not see at all in dim light, or only after long adaptation, *hemeralopia* or *night blindness* exists (retinitis

pigmentosa; chorioiditis diffusa; glaucoma; retinal detachment; general disturbances of nutrition).

By *photopsia* is meant an abnormal sensation of color or light due to retinal lesions, or to central irritation, by *metamorphopsia*, the distorted vision, occurring in lesions of the retina and chorioid, in which straight lines look curved or jagged, or letters seem out of place. The term *macropsia* is used to designate the symptoms "of seeing things too large" (e. g., strained accommodation), while *micropsia* is the term for "seeing things too small" (e. g., paresis of accommodation).

In many functional nervous cases a study of anomalies of refraction has an important bearing upon etiology and treatment. We distinguish the normally refracting eye (*emmetropia*) from the short-sighted eye (*myopia*) and the far-sighted eye (*hyperopia*, *hypermetropia*). When in order to secure normal visual acuity it is necessary to use not simple spherical lenses, but lenses which correct certain meridians only, the condition of *astigmatism* exists; if the curvature in each meridian is equal throughout its course, but the curvatures of different meridians vary so that the meridians of greatest and least curvature are approximately at right angles to each other the astigmatism is *regular*; when the vertical meridian has the greatest curvature it is astigmatism *with the rule*, but when the horizontal meridian has the greater it is astigmatism *against the rule*. The astigmatism is *irregular* either when the meridians of greatest or least curvature are not approximately at right angles or when the curvature in a single meridian is irregular.

Anomalies of vision due to *disturbances of accommodation and convergence* may also be mentioned here, although they belong more properly to disturbances of motility. If near vision is faulty, or cannot be maintained long, *asthenopia* exists. If this is due to faulty power of accommodation, it is known as *accommodative asthenopia*. The most frequent form of this is due to the progressive sclerosis of the crystalline lens, which normally diminishes the power of accommodation as life advances, and soon after the fortieth year the near-point of a normal (emmetropic) eye comes to lie more than 25 cm. distant (*presbyopia*). Accommodative asthenopia due to paralysis of the M. ciliaris often follows diphtheria.

Another form of defective near vision is *muscular asthenopia*, especially that due to faulty convergence (most often seen in uncorrected myopia) with consequent faulty binocular near vision (*convergence asthenopia*).

The lesions of the eye, most important for the neurologist, discoverable by the *ophthalmoscope*, may be briefly referred to here. In optic neuritis the papilla looks turbid, grayish-red in color, its margins are indistinct, and there are often minute hemorrhages in or near it and the veins may be tortuous and dilated. It is most often due to cerebral disease, especially when bilateral. If the papilla be swollen (2 diopters or more) and enlarged, we speak of *choked disk*; it is most often due to increase of intracranial pressure (brain abscess, hydrocephalus, syphilis, tumor).<sup>1</sup> *Unilateral optic neuritis* occurs in diseases of the orbit, in various infections and intoxications, and in beginning tumor, etc. Optic neuritis

<sup>1</sup> See Cushing and Bordley, *Johns Hopkins Hosp. Bull.*, 1909, xx, 95.

may be followed by optic atrophy; the papilla has indistinct margins, but becomes whiter as the swelling goes down; the arteries gradually narrow and have white sheaths (*neuritic optic atrophy*).

*Retrobulbar neuritis* occurs in two forms—acute and chronic. The acute form is seen in multiple sclerosis, the chronic in diabetes, in various intoxications (tobacco, alcohol), and sometimes when its cause is not known. Here the ophthalmoscopic picture is very different from that described above for “optic neuritis;” no distinct alteration can be seen in the papilla N. optici for some time, although after a while a retrograde atrophy leads to pallor, especially on the temporal side of the disk.

When the papilla N. optici looks white, grayish-white, or greenish-white instead of its normal rose color there is atrophy of the optic nerve (*atrophia N. optici*); ophthalmologists distinguish several types: (a) Simple, (b) retinal, (c) glaucomatous, and (d) neuritic atrophy.

Of *simple optic atrophy* in which there is simply pallor of the disk with retention of its well-outlined margins and but little change in the vessels, three main types are described: (1) *That occurring in tabes and dementia paralytica*, the changes in the disk being visible from the beginning of the visual disturbance; (2) *retrograde atrophy*, following upon diseases at base of brain, *e. g.*, tumors, chiasm lesions, sphenoidal-sinus disease, etc., the changes in the papilla becoming visible through the ophthalmoscope only after the visual disturbance has lasted some time; and (3) *atrophy after occlusion of the A. centralis retinæ*, in which the simple atrophy differs from (1) and (2) by the existence of a high grade of vascular constriction.

The so-called *retinal atrophy of the optic nerve* is characterized by a yellowish-white turbid papilla, constricted bloodvessels, and degeneration of the peripheral retina; it occurs in diffuse degenerations of the retina (chorioido-retinitis; retinitis pigmentosa).

The form of *atrophy accompanying chronic glaucoma* is very characteristic. The increased intra-ocular pressure leads to “glaucomatous excavation,” and this after a time is followed by atrophy with abrupt kinking of the bloodvessels at the margin of the papilla.

**Anomalies of Sense Perception Known as Illusions, Hallucinations, and Pseudo-hallucinations.**—Normal *sense perception* is more complex than mere sense impression; it involves *association* and *assimilation*. An “object” is composed only in small part of sense impressions directly received by way of the peripheral sense organ, but very largely of revivals of impressions earlier experienced (*reproduced images; representations*). A *percept* arises largely through the arousal of a host of old experiences by a few newly entering sensations; perception is not, as it may seem, the mere entrance of a group of new sensations. The percept as thus constituted depends, especially in its meaning or the value which it has in consciousness, upon the general direction of thought or upon the most outstanding idea in the immediately preceding moment; this is often, though inadequately, expressed by the term *attention*.

We can profitably pass on to the consideration of the anomalies of sense perception known as *sense deceptions*, including illusions, hallucinations, and pseudo-hallucinations. In all these sense deceptions there is an

"abnormal relation between the objective and the subjective setting, and the factors that control the course of the associations are not suited to the circumstances of the physical world." Events occur in accord with the laws governing normal perception, but the conditions are unusual and individual, not those which obtain with the mass of healthy people.

By an *illusion* we mean a sense perception in which the subjective factor takes too large a part in the process of assimilation, so that the mental supplementation of the sense impressions entering into it leads to a result contradictory to the reality as determined in more accurate examination with the same sense organ or on correction by control through other senses.

By an *hallucination* we mean a sense perception in which the abnormal relation between subjective and objective conditions is very pronounced; objective sense impressions may be entirely absent or present in unusually small amount and dissimilar to the content of the hallucination, and yet the sense perception is vivid, possessing for the patient the ear-marks of objective reality, being definitely localized in perceived space; the annihilation of the perception normally localized in this space is unexplained.

By a *pseudo-hallucination* we mean a psychic experience which resembles a sense perception in intensity and in its involuntary appearance (thus, different from a mere image, idea, or representation called up by association), but differing from a normal sense perception, from an illusion, and from an hallucination in that the patient notices the absence of the character of objectivity.

We have then a series of transitionally related psychic phenomena beginning with sense impressions at one end and pure reproductions at the other, the series consisting of (1) sense impressions, (2) normal sense perceptions, (3) illusions, (4) hallucinations, (5) pseudo-hallucinations, and (6) pure reproductions (ideas, images, representations). In normal people in ordinary circumstances, the sense deceptions (illusions, hallucinations, and pseudo-hallucinations) do not occur; they are experienced occasionally by normal people in extraordinary circumstances and frequently in mentally abnormal people in ordinary circumstances.<sup>1</sup>

**Illusions.**—The psychic experiences known as *illusions* are best studied in connection with the sense of sight and hearing, since with these senses one can be sure as to the character of the external stimuli acting. Sounds heard and sights seen are misinterpreted owing to the incorrect supplementation (by reproductions—images; ideas) of the sense impressions received. A melancholic patient, hearing the whistle of a locomotive engine, says that she hears the cry of her child being murdered, or, seeing a relative, fails to identify him as such, but takes him to be a stranger and impostor. Even normal people are often the subjects of illusion when the sense impressions are feeble and the mind is in a state of expect-

<sup>1</sup> Some of the more important contributions to the general bibliography of sense deceptions are the following: Kahlbaum, *Die Sinnesdelirien Allg. Ztschr. f. Psychiat.*, xxiii, 1-86; Parish, E., *Hallucinations and Illusions*, 1897; Sully, J., *Illusions*, 1883; *Text-books on Psychiatry* by Krafft-Ebing, Kraepelin, and Paton have good chapters on the subject. The discussion in Störing's volume is excellent.

tation; for example, an anxious mother may take the mewing of a cat for the cry of her sick child in a distant room. But the stronger and more distinct the sense impressions when illusions occur, other things being equal, the greater the degree of abnormality indicated. Deficient attention to sense impressions (as in alcoholic delirium) and short duration of the sense impressions favor the origin of illusions. Tense expectation, emotional states or moods, a lively imagination, or pathologically increased facility of reproducing ideas will also favor illusion, for in these circumstances certain ideas are prone to be held focal in consciousness and they will obviously have the best opportunity for coöperating with any sense impression which may enter. In insanity, illusions are probably just as important as hallucinations, the sense deceptions being due to the delusional ideas which are in consciousness at the time of entrance of the sense impressions.

**Hallucinations.**—These may be elementary or complex. An *elementary hallucination* is one in which the subjective experience approaches that of a simple sense impression (*e. g.*, a flash of light, a simple sound), while a *complex hallucination* is one in which the subjective experience approaches that of a compound sense perception (*e. g.*, vision, voice). Hallucinations and illusions conform to the law of excentric projection or exteriorization of sense perception; the patient “projects” what he sees, hears, or feels into the external visual, auditory, or tactile fields, or localizes the sensation either in space outside or in various parts of his body.

Hallucinations seem even more real to those who experience them than do normal sense perceptions; their power is irresistible; the patient is compelled to believe in them, the judgment being quickly overcome by them. This peculiar feature is due in part to the sensory distinctness, the seeming objective reality of the experience, but much more to the fact that hallucinations most often correspond to the pathological ideas of the patient, to his abnormal thoughts, fears, or wishes. Thus, although sense deceptions often play a part in the origin of delusions, some persistent direction of thought (*Aufgabe*) is still more often provocative of sense deceptions. Further, the strong feelings and emotions co-existent with the hallucinations go far toward giving the latter their power in the mental life.

Among *visual hallucinations* the elementary types are represented by flashes of light or color, stars, flames, and the like, the complex types by *visions* in which, especially at night, illuminated animal and human forms appear, or God, angels, the Devil, or various horrible shapes may be seen; if the hallucination occur in the daytime, which is less common, the patients complain of seeing a black dog, the face of a corpse looking in at the window, and the like. Sometimes in hemianopsia the visions are projected to the blind halves of the visual fields; occasionally, as in convalescence from alcohol delirium, visions which have begun to disappear may be recalled by pressure on the closed eyes (Liepmann).

Even more influential in mental life than visual are *auditory hallucinations*, the elementary forms of which are called *akoasmata*, the complex forms being known as *phonisms* or *voices*. The *akoasmata* consist of indefinite noises of different sorts—rustling, murmuring, thundering, and the

like; the phonisms or voices are often whispered words, more rarely words or brief sentences spoken aloud. The voices make unpleasant references to the patient, they insult or threaten him ("rascal," "thief"). They may emanate from the patient's children, who are being murdered by an enemy, from an annoying neighbor, or it may be the Deity or Satan who addresses him. Occasionally a voice may bring happiness and inspiration, but this is unusual; nearly always the voices irritate and pain, arouse suspicion and defence, cause anxiety or confusion; they may command, and the patient may be led to the most unnatural acts in the belief that he is obeying the order of a supernatural power. The patients can tell the distance and direction from which the voices come—they localize them in space—a fact closely related to the objectivity (perception value) which attaches to the voices. If the voices are heard in one ear only, there is usually abnormal stimulation of the auditory path on that side, and the hallucination (or illusion) can often be called forth by electric stimulation.

A word must be said about *audible thinking* (*Gedankenlautwerden* of the Germans), not of the delusional form of it in which the patient is convinced that his thoughts are audible to other people, although he does not hear them himself, but of the hallucinatory form in which a man hears his own thoughts, perhaps in his feet or in the ticking of a clock or the ringing of the doorbell. The patient hears the voices only when he pays attention to them. Sometimes a patient's thoughts seem doubly existent to him, first, in the normal way and then also as audible voices (*double thinking*); sometimes the voice dictates what the patient writes or it may repeat what the patient reads; occasionally the voice precedes the reading. Many of these "inner voices" are examples of pseudo-hallucination rather than of true hallucination.

It is extremely difficult to distinguish *olfactory* and *gustatory hallucinations* from illusions of these senses, and the same is true of *hallucinations of visceral sense*. They are, however, of great practical importance, for they often account for the refusal of food. If the food smells of feces or foul sweat, one can scarcely blame a patient for not eating it. In the domain of the cutaneous senses, *tactile* or *hepatic hallucinations* and *thermal hallucinations* are not infrequently met with, the former especially in toxic psychoses; some of the subjective dysesthesias may be hallucinatory in nature, as may also certain so-called *psychalgias* or *topoalgias* (*hallucinations of pain sense*). Much study has been devoted also to *hallucinations of the movement and position senses*, sometimes referred to as *kinesthetic hallucinations*. They lead to the maintenance of peculiar attitudes and also affect the voluntary movements, being sometimes the cause of imperative positions or monotonous defensive movements. The hallucinations may concern the eye-muscle movements, the locomotory movements, or the speech-muscle movements. Sometimes there are *hallucinations of vestibular sense*, the patient having the sensation of progressive or rotary movement. Doubtless many impulsive acts and various psychomotor inhibitions are traceable to hallucinations as a cause; in epilepsy and alcoholism there may be an irregular *hallucinatory agitation*, sometimes combined with paroxysms of rage.

While the hallucinations above described are subjectively equivalent to perceptions, the patients believing that they are dealing with something objective, this is not true of what Hagen has called *pseudo-hallucinations* (psychic hallucinations of Baillarger; apperception hallucinations of Kahlbaum; imagination deceptions of Kraepelin). Faces printed pages, groups of people appear for a moment and vanish, to be followed by others not logically connected with them. They seem to stand before the eyes, but are not at all related to the black field of vision of the closed eyes. Despite their sharp contours and lively colors, the images do not possess the character of objectivity; the patient says he sees them with "inner eyes," not with his "outer eyes." Although the objects seen "stand before the eyes," they are not related to any real objects about him. In pseudo-hallucinations of hearing the patients complain that they are "forced to hear internally" (not with their ears) things which they least wish to hear. Obviously, although these psychic states resemble perceptions on the one hand, and lively reproductions of mere ideas on the other, they differ from both. They differ from percepts in that the patients recognize the lack of objective reality (physical) and their independence of the movements of the peripheral sense organ concerned; they differ from ordinary reproductions (images, ideas) in that they have greater intensity and are independent of the will.

#### DISTURBANCES OF MORE COMPLEX PSYCHIC PROCESSES.

**Anomalies of Consciousness as a Whole.**—Consciousness is the fact that we have experiences—sensations, ideas, feelings, etc. To understand consciousness, therefore, we must understand experience fully. In spite of ages of speculation we are certainly, as yet, only making a beginning in the understanding of experience, in that we are gradually determining some of the conditions under which it occurs. These conditions are (1) partly of the order of physical facts (*e. g.*, light-waves, sound waves); (2) partly of the order of physiological facts (*e. g.*, processes in sense organs, nerves, and central nervous structures); and (3) partly of the mind itself (*e. g.*, imagination, feeling, volition).

As ideas come and go in consciousness we are aware that some of them are more prominent or outstanding than others. We commonly speak of *paying attention* to these more prominent ideas. Subjectively this prominence of certain ideas is, at times, definitely accompanied by a peculiar feeling of "activity," a sense of strain or effort, which is in contrast to the feeling of "submission" (passivity) which accompanies a distracting sensation or memory. When ideas have the prominence just referred to we feel that they are *ours* in a peculiar sense—that we are *doing* something with them. In analogy with the visual field, we often refer to these prominent ideas as being in the focal part of consciousness (*Apperception* of Wundt), while the other ideas are marginal (*Perception* of Wundt).

By the *mind* we usually mean not simply the abstract *state of consciousness* or the *total content* or *field of consciousness* at any given moment; we include in it also that unity with past experience through which the

present moment has a meaning and value, dependent on the whole education of the individual which makes it truly personal. When we distinguish between the *activity of the mind* (Bewusstseinstätigkeit) and the *content of the mind* (Bewusstseinsinhalt) we should recognize that an abstraction is being made between two aspects of every idea which have no significance apart from one another.

In the central neurones changes take place which permit the calling up of memory pictures in simple and complex groups. The totality of memory pictures possibly revivable is what is designated as the *contents of the mind*. This totality of possible memories is divisible into three parts: (1) That which refers to the *external world*; (2) that which refers to the *body* or *soma*; and (3) that which refers to the *personality* of the individual. Wernicke has given to these three parts of the mind's contents the names *allopsyche*, *somatopsyche*, and *autopsyche*.

The *allopsyche*, or portion of the mind's contents which concerns the external world, is made up of memories of concrete things, the result of sense impressions, their combination into perceptions, and the shorthand epitomes of these which we call concepts and notions. The *allopsyche* of an individual will depend then largely upon his sense organs; that of the congenitally blind will differ greatly from that of the congenitally deaf, and that of the anosmic will be wholly other than that of the ageusic. Given brains of equal birth, the *allopsychic* contents will vary quantitatively according as the bearer is a resident of a city or a backwoodsman, or may differ qualitatively with the environment (*e. g.*, tropical or arctic).

The *somatopsyche* or portion of the mind's contents which concerns the body consists of all the memories which represent residues of previous visceral, muscular, fascial, and cutaneous sensations. In all probability the so-called "local sign" which accompanies the exterior sensations is to be counted as a part of the body consciousness, the local sign of a visual sensation representing, for example, an "organ sensation" of the retina (Wernicke). Here the *allopsyche* and the *somatopsyche* overlap. The latter depends largely upon the condition of the viscera, and the tendency is to relate the hypochondriacal symptoms of mental disease to anomalies of visceral sensations.

It is probable that in every *allopsychic* sense perception the *somatopsyche* plays a part; indeed, this *somatopsychic* participation may be the main difference between a perception and a revived image of memory, for in the perception an organ sensation (local sign, feeling tone) is present while it is absent in the memory pictures (Wernicke).

The various projection fields of the *somatopsyche* must be intimately connected with one another by associative neurone systems. The connections of the memory pictures concerning the body are much more intimate than those uniting the memories of concrete things of the external world on account of the unalterable relations of the different parts of the body as contrasted with the continuous changes in the world outside the body. In sleep the impressions from the external world cease to affect the brain, although bodily impressions are still entering it. The external world is at times cut off; the body is always there. When the body is undergoing marked changes, as at puberty, the climac-



teric period, there must be a marked alteration in the character of the afferent impulses from the viscera. Is it any wonder, then, that disturbances of somatopsychic consciousness tend to prevail at such periods?

The relative constancy of bodily sensations, the continuity of the somatopsyché, as compared with the variety of sensations from the external world and its discontinuity, give rise in each individual to a conception of unity, of individuality, of the body consciousness (*primary ego* of Meynert).

The *autopsyché* or portion of the mind's contents which concerns the personality or individuality is something built upon the *allopsyché* and the somatopsyché which serve as its foundations. The *autopsyché* or *consciousness of personality* includes all the mental possessions and acquisitions resulting from instruction, culture, and education, the influence of the family and of the social milieu. Speech and written language play an important part in its development. School knowledge is among the early acquisitions of the *autopsyché*; later on come all the experiences peculiar to the individual. The interest in work, in family, in friends, and in other people belongs here. The development of the *autopsyché* depends largely upon the kinds of *allopsyché* and somatopsyché which form its foundations; as Wernicke puts it, "consciousness of personality is a function of the consciousness of the external world and of that of the body. . . . The man with sharp senses and strong body must develop a mental personality different from that of a man with dull senses and a weak body." An individual's own estimate of his place in human society depends upon the state of his *autopsyché*. Among the striking anomalies in this domain may be mentioned ideas of grandeur or of self-depreciation, ideas of reference and delusions of persecution.

There seems to be no doubt that the somatopsyché, the *allopsyché*, and the *autopsyché* are, to a certain degree at any rate, localizable; or, more exactly, their physiological correlates appear to be to some degree localizable in the cerebrum. Clinical and pathological studies of dementia paralytica, of cerebral arteriosclerosis, etc., show us how the *autopsyché*, the *allopsyché*, and the somatopsyché may separately or successively become involved.

**Circumspection; Confusion; Unconsciousness.**—In the normal awake state there exists a certain equilibrium in perceptions, ideas, and feelings, the external manifestation of which is known as *circumspection* or *presence of mind* (*Besonnenheit* of German writers); here the mind is calm and collected with its faculties ready at command. It is the opposite of *mental confusion* (*Verwirrtheit* of German writers) or *incoherence*, in which ideas are jumbled and chaotic, the mind perplexed or bewildered, the speech aimless or unintelligible, the acts disconcerted. By *unconsciousness* is meant a state in which the psychological powers are wholly in abeyance (dreamless sleep, profound narcosis). Attempts have been made to distinguish various grades of pronounced lowering of consciousness, and references will be found to a series of states designated in order as *somnolence*, *sopor*, *coma*, and *carus*.

**States of Psychical Cloudiness or of Clouded Consciousness; Mental Fog; Twilight States; Automatism.**—By *mental fog*, *psychical cloudiness*, or a *twilight state* (*Dämmerzustände* of German writers) is meant a condition in which there is diminution of clearness of consciousness or a loss of memory for a part of past experiences or both together. Such mental fog is often met with in epilepsy and in hysteria. In *epileptic mental fog* there are often fluctuations of consciousness intensity, partial loss of power of recalling the past, alterations of mood and false identification. In *hysterical mental fog* we meet the so-called *monoideic* and *polyideic somnambulisms* and *fugues*. In somnambulism an individual thinks and acts in an odd way, different from that of other people, while he is in some way like a person asleep. During a *monoideic somnambulism* the patient acts as though in a dream, but the phenomena of the dream are most perfectly and intensely developed.

In *fugues* (*flights* or *ambulatory automatisms*) we have to deal with the hysterical mania for running away. During the abnormal state a certain idea or trains of ideas develop to an exaggerated degree (attempt to elude capture; desire for travel, etc.), and lead to the flight. At the same time other thoughts which would normally counteract this idea or train of ideas appear to be suppressed; the patient may forget his name, his family, his social position, or, in other words, his ordinary personality. After the fugue is over and circumspection returns the patient has forgotten many or even all of the events of the flight, and also the ideas which dominated him during it. Through hypnosis it is sometimes possible to reproduce the mental state of the fugue artificially.

Intermediate between fugues and monoideic somnambulisms are the so-called *polyideic somnambulisms*. These are characterized by the multiplicity of the ideas that fill them. Instead of the one idea or event of the monoideic type, we see evidence of the existence of a series of ideas succeeding one another in the somnambulant state, ideas which pertain to some strong feeling which develops independently of the rest of consciousness. Where still larger systems of ideas and feelings develop independently we get the manifestation of what are known as *double personalities* or *multiple personalities*.

Twilight states are usually temporary, but they may last for varying lengths of time; they may last for weeks or months. The cessation of the state is, as a rule, sudden, the patient waking as though from sleep. Indeed, sleep seems to be the physiological paradigm of a twilight state, dreams being the analogue of the psychical activity, but differing from the consciousness of the twilight state in that usually they are unaccompanied by motor reactions.

**Disorientation.**—Many patients are not well oriented as to their own persons and their spatial and temporal relations. For differential diagnostic purposes it is very important to determine the presence or absence of certain other symptoms at the same time orientation is tested, especially anomalies of mood, ideas of persecution, and hallucinations. A few simple questions along with those concerning orientation may quickly give the clue to the existence of a hallucinatory confusion, of a paranoid state, or of some other well characterized type of mental disorder. Especially

important for the general practitioner is the valuation of the syndrome in which disorientation as regards time and space is combined with so-called pseudo-remiscences, in which the patient narrates long tales of recent events as though perfectly true, which on inquiry are found to have no basis in fact (*Korsakow's syndrome, polyneuritic psychosis*).

**Anomalies of Attention.**—The importance of disorders of attention in psychiatry is coming to be very generally recognized. The power of directing thought toward a definite task (*Aufgabe*) (*vigility*), and of maintaining this task despite intercurrent stimuli (*tenacity*) are essential in all intellectual operations. We meet with most variable ability to direct and maintain the task, some being able to do this easily, while others are easily distracted. The greater the distractibility the more dirigible from without, the less thorough is the intellectual work. The term *hyperprosexia* is by some reserved for the pathological persistence of a task in which the mind is concentrated upon certain abnormal ideas or sensations (hypertenacity), while *aprosexia* is defined as a state in which there is inability to set or to maintain a task (*e. g.*, in imbecility). There is both hypovigility and hypotenacity. Some writers use the word *hyperprosexia* to indicate a state in which there is increased tendency to change the task set (*hypervigility*) with diminished tenacity.

**Examination of the Recording Faculty.**—For testing the *recording faculty*, which depends in large part upon attention, much simpler methods must be used in medical practice than in experimental psychological examinations, and we have to thank Ranschburg<sup>1</sup> and Boldt<sup>2</sup> for supplying us with such simple methods. It is necessary to test the recording faculty in several different domains, and most experiments consist of acoustic and optic tests. Thus, in one group of experiments the acoustic word-memory, using the principle of idea associations, may be tested. The patient memorizes fifteen pairs of words, of which five represent customary word compounds, like dog-kennel chair-leg; five according to the laws of causality (spatial and temporal coincidence), like fish, pond; day, night, etc.; . and five according to the principle of similarity of sound, like wound, wonder; die, dynasty. In another group of tests the memory of persons is tried by using twenty-five portraits, from which the patient is to recognize five. In a third group the memory for colors may be tested. Many colored skeins of red, blue, green, and yellow wool are shown and a definite example of each is to be recognized. In a fourth group the orientation memory is tested. One shows the patient a chart containing six simple geometrical figures, exposes it for twenty seconds to permit of certain perception, and after a time the patient is required to pick out these six figures from a chart containing twenty or twenty-two figures. In a fifth group the memory for nonsense words is tested, five pairs of words being chosen, *e. g.*, lum, rar; grat, smor. In a sixth group of tests the memory for names is tried. The patient is shown five photographs and the name and surname given. Later on the patient picks out these five photographs from a group of twenty-five and tries to give correct name and surname. Lastly, in a seventh group the

<sup>1</sup> *Monatsschr. f. Psychiat. u. Neurol.*, Berlin, 1901, ix, 241 to 259.

<sup>2</sup> *Ibid.*, 1905, xvii, 97 to 115.

memory for numbers is tested. Some number is given to each one of the five fingers, and later on, naming each finger, he is asked to give the corresponding number. The reproduction is best tried shortly after the perception (five or fifteen minutes) and again after twenty-four hours.

The recording faculty consists of two factors, namely, perception and reproduction. Of course, where perception is deficient there can be no reproduction later. One makes sure, first, that the task is correctly understood. The patient is warned not to try to reproduce repeatedly in his mind the things perceived, for even in normal persons the test turns out less favorably later when such efforts are made. The first reproduction is permitted after five minutes have elapsed, the second after fifteen minutes, and twenty-four hours later the third test is made. The reproduction at the end of twenty-four hours may in normal persons be much better than that at the end of five minutes, but in patients with injured recording faculty this is not the case. The recording faculty appears to be the one of the mental powers first and most seriously to suffer in dementing processes. Injury to the recording faculty as manifested in loss of memory for most recent events never exists long alone; there are added to it very quickly other characteristic symptoms of mental deterioration. The recording faculty is most efficient in childhood especially between the twelfth and the fourteenth years of life.

When the recording faculty begins to be impaired it seems to be the memory for numbers which goes first. As Kraepelin has emphasized, studies of this recording faculty have drawn the attention of clinicians to a group of anomalies previously entirely unknown to us, viz., (1) the quick fading of perceptions, and (2) the slow development of perceptions, both of which appear especially to favor the origin of memory falsifications.

**Anomalies of Ideation.**—Great importance attaches to the examination of the processes of ideation; that is, to the formation and association of ideas. Ideation may be disturbed on the formal side, or there may be falsification of the content of ideas. Of the formal disturbances of association may be mentioned: (1) Anomalies in velocity; (2) anomalies in the order of association or mode of reaction; (3) anomalies in the intensity and duration of ideas; (4) anomalies in comprehension or apperception; (5) anomalies in so-called identical reproduction (memory); (6) anomalies in so-called original production (phantasy). The falsification of content of ideas is manifest in the production of the so-called insane ideas or delusions.

**Anomalies in the Velocity of Association; Flight of Ideas; Thought Inhibitions; Stupors.**—The association of ideas may be pathologically accelerated or pathologically retarded. Even in health there seems to be an *acceleration* of the velocity of association during excitement, especially during joyful excitement. In maniacal states we meet with a peculiar form of association known as *flight of ideas* (*Ideenflucht* of the Germans), which is often associated with the form of motor agitation known as *pressure of activity* and accompanied by marked gesticulation and rapid speech (*logorrhæa*). In a careful study of the flight of ideas, however, it is found that we have to deal less with acceleration of association than

with an alteration in the mode of reaction. The quicker thinking of the maniacal patient seems rather due to the fact that in the same time unit more different objects pass through consciousness than in normal states (Liepmann). In the healthy person ideation is dominated by some principal idea, while in mania the process of association is not controlled by such a dominant idea. One of the most striking characters of the associations in the so-called flight of ideas is the frequent *change of direction of ideation*.

In normal man the kind of reproduction of ideas and the duration depend not only upon the reproduction tendency present in the individual, but also upon the effect of *concentration upon the task set*. Consciousness is, in a way, set, determined, or prepared in the sense of a specific task. In mania the task set has but little effect upon the process of association as compared with healthy states. The healthy man goes only very gradually from one ideational direction to another and tends to remain for a longer time within the confines of the idea made dominant by the task set. The healthy man, it is true, may for a short time experience rapid associations frequently changing in direction,\*the quality of the association then being superficial, like those seen in flight of ideas; but he is incapable of so continuing to associate for any length of time, whereas the characteristic of flight of ideas is the continuance of such associations until the physical strength is exhausted.

At bottom the disturbance known as flight of ideas must be largely a disturbance of attention. The main change in the attention seems to be an increase in the capacity for directing the thought quickly and energetically to single ideas (hypervigility), and also away from them again (hypotenacity). To explain the abundance of ideas reproduced, however, it is necessary to assume also an alteration of the power of reproduction, permitting the appearance in consciousness of a wealth of lively ideas capable of being easily replaced one by another (Isserlin).

Psychiatrists distinguish a *primary flight of ideas* (e. g., reminiscence flight in neurasthenic states and in the exaltation of mania) from a *secondary flight of ideas* due to quickly changing hallucinations and delusions; in the latter case the idea flight is not a new disease symptom, but is secondary to the affective excitation called forth by the hallucinations or delusions. When the associations run unbridled by a dominant idea, as they do in the marked forms of idea flight, there is an inability to follow anything like a normal train of thought (*secondary incoherence*).

Pathological *retardation* of association (*thought inhibition*) may also appear, either as a primary phenomena or as a secondary symptom. When pronounced it leads to the condition known as *stupor*. Such slowed association is nearly always associated also with *motor inhibition* in which the so-called voluntary movements are slowed or absent altogether. The patient is unable to complete the series of association between the initial idea and the terminal motor idea. Speech becomes more and more slowed until there may be complete mutism. The various voluntary muscles of the body may be held rigidly (as in catatonic rigidity)

As examples of *primary stupor* may be mentioned: (1) the so-called excitation stupor which appears in the slowly developing conditions of

cerebral excitation, and (2) the stupors which appear in the course of various intoxications. Unless the history is known in such cases it is sometimes difficult to tell whether one is dealing merely with an inhibitory process or with actual defect (dementia). As *secondary stupor* may be designated the form which accompanies or follows various states of psychic depression. An emotional shock or a paroxysmal anxiety condition may call forth a severe secondary thought inhibition. A special form of secondary stupor due to delusions and hallucinations accompanied by strong affective states is met with in certain conditions of catatonic rigidity (pseudo-stupor of Westphal).

The so-called waxy flexibility (*flexibilitas cerea*) may be regarded as a part of the general inhibition of association.

**Anomalies in the Order of Association or Mode of Reaction.**—Perhaps most progress in recent years has been made in the study of the *mode of reaction* in association-tests. A study of diseased conditions reveals marked deviations from the normal modes of reaction. It is customary to classify the kinds of association as follows:

A. Sense of test words correctly understood.

(a) Internal associations. (1) Associations of coördination and subordination. (2) Predicative associations. (3) Causality associations.

(b) External associations. (1) Associations of spatial and temporal co-existence. (2) Identities. (3) Speech reminiscences.

B. Sense of test words not understood.

(c) Test words acting only through their sound. (1) Word supplements. (2) Sound associations and rhyme associations. (A) Intelligible. (B) Nonsensical.

(d) Test words acting only by setting free reactions. (1) Repetition of test word. (2) Repetition of earlier reactions without sense. (3) Associations for words used earlier. (4) Reactions without recognizable connection.

It is also important to note how many of the reactions are more *objective* in nature and how many of them are more *subjective* (*egocentric*).

A word of explanation as to the meaning of some of the terms used above seems desirable. By an *internal association* is meant one directed toward the sense of the test word, while an *external association* is one dependent upon custom, habit of speech, and sound relation. In the *predicative associations* there is some affirmation or assertion regarding the object of the test word. In the *causality associations* there is a relation of cause and effect. By *word supplementation* one means only such reaction words as, taken with the test word, make together an indivisible word. Such reactions belong to *clang reactions* in the wider sense.

A distinction is made between *mediate association* and immediate (Aschaffenburg). In the former the only way one can understand a connection between the test word and the reaction is by assuming some intermediate member of an association series. As a rule, such intermediate member turns out to be a clang association for the test word,

while the relations of the reaction word to the intermediate member may be any one of the forms of association mentioned.

It is customary to designate instances in which more than sixty seconds elapse before an answer is given to the test word as "faults." Reactions in which there is simply a repetition of the test word are in a class by themselves. It is well also to count up the number of perseverations and repetitions which occur. By *perseveration* is meant a reaction in which there has been evidently a connection on an earlier test or reaction word instead of to the actual test word given; by *repetition* is meant a reaction in which the test or reaction word immediately preceding recurs.

*Discursive associations* should also be especially noted (*Weiterschweifen* of Isserlin). By this is meant a spontaneous continuation of the association. Such further association may be intelligible or nonsensical.

Special attention should be paid to the instances in which the test word is repeated by the patient. When this is a marked feature in a case it should be determined whether it is due to anomalies of comprehension from marked distractibility, in which the attempt to concentrate leads to repetitions, or whether it is dependent upon inability to draw upon the reproductive elements sufficiently, that is to say, is due to a lack of associated ideas (as in the thought inhibition of depressive states). A patient will often give the clue himself, since he may mention, on the one hand, that nothing occurs to him, or, on the other, that so many things pass through his mind that he repeated the test word in order to answer correctly. The two types are further distinguishable by the fact that, in the one, reproductions often in large number appear immediately after the repeated test word, while in the instances due to thought inhibition associations occur only after quite a pause, and often only after several slow repetitions of the test word.

Experience with these association tests soon permits one by their aid to differentiate quickly among the great groups of psychotic symptoms. Another advantage of systematic monthly association tests consists in the ability to follow the exacerbation or retrogression of symptoms.

In valuing the results of association tests one should familiarize himself, first, with a large material obtained from healthy people. Educated people show, on the average, a more "superficial" type of reaction than uneducated people, the latter reacting more to the sense of the test word than the former. Differences in individuality determine also certain important variations in association. Jung and Riklin distinguish two main types: (1) *Objective types*, and (2) *subjective types with egocentric adjustment*. To the objective types belong those persons by whom the test word is understood objectively, either according to its sense or as a speech stimulus, while to the subjective types belong (a) the so-called "*constellation*" types, in which personal experiences (idea complexes) with strong feeling tone appear in the associations, and (b) the "*predicate*" types, which present lively, subjectively valued ideas.

The reaction time of normal persons varies greatly according to the age, education, and individuality. The more phlegmatic and the intellectually feebler individuals show, as a rule, longer reaction times. The quality of the association is an important factor in its duration; thus the

more complicated an association is psychologically the longer the reaction time. The associations which are most used in daily life are those with the shorter reaction times, a fact which explains the velocity of "external" associations as contrasted with "internal" associations. One of the most important results of association tests is the bringing of the proof of the delay in reaction time in associations to which *idea complexes with strong feeling tone* are attached.

Association tests in the *melancholic states of manic-depressive insanity* reveal a marked prolongation of the reaction time and apparently a characteristic change in the content of the associations. There appears to be a limitation to idea change. The reaction type is usually predominantly objective. Even in the very delayed associations when the reaction is recorded, it, as a rule, corresponds in sense to the test word.

In the *maniacal states of manic-depressive insanity* the striking features are a "flattening" of the associations and the replacement of associations due to the sense of the test words by associations which depend upon custom, speech, and mere sound relations (clang associations). The frequency of discursive associations (*Weiterschweifen*) in mania has been pointed out by Isserlin. As in flight of ideas in general the reaction times are not shorter than normal; indeed, they are sometimes really longer, although in maniacal states one does meet perhaps with a greater number of short reaction times than in health. The most characteristic phenomenon in mania is the greater number of single ideas produced during a given time—many more than one ever meets with in health. It is the extraordinarily frequent *change of direction* of the ideas which is so striking. Healthy persons are incapable of associating so quickly with such frequent change of direction and in such a "superficial" manner as is characteristic of the idea flight of maniacal states.

Progress has been made in the study of associations in the so-called *mixed states of exaltation and depression*, in which certain of the signs of mania are associated with certain of the signs of depression. One tries to find out whether the reaction time is prompt or delayed, whether the type of reaction is according to the sense of the test words or superficial, whether the direction of the ideas is changed frequently or rarely, whether there are many or few perseverations and repetitions, whether the form of the answers indicates a precise reaction or a vague "talking around the subject," whether the feeling tone is positive or negative, and whether or not egocentric relations, faults, and test-word repetitions are prominent (Isserlin). In the "mixed states," egocentric relations may be extremely frequent. Test-word repetitions tend also to be especially common in these mixed states.

In view of the help which comes from a study of the result of association tests, some physicians seem inclined to overestimate their value. It should be borne in mind that they reflect only one side of mental life, and that one must not neglect other psychological methods of examination. The general practitioner has not yet formed the habit, however, of resorting to association tests, and to him the method can be heartily recommended as a means of extending the anamnesis. Especially in the differential diagnosis of manic-depressive insanity from dementia precox



the method will be found helpful. If one bears in mind the characteristic signs of inhibition and excitation revealed in the patient's *entrance upon the task* of association in manic-depressive insanity, and compares therewith the refractory, perverse behavior with regard to the task characteristic of the patient suffering from dementia precox, and observes, further, the stereotyped replies and repetitions or perseverations which may seem to disregard the test words, the paralogia, the mannerisms which come out during the association tests, one will find the method one of the best available in distinguishing these two types of psychosis from one another. Even in cases where strong inhibition simulates apathy and dementia the association experiment helps to differentiate. If, in spite of long reaction times, we see an effort to understand the sense of the test word, we can, as Isserlin emphasizes, make the diagnosis of manic-depressive insanity even when the change of direction of the ideas is only moderate and there seems to be relatively little affectivity. In dementia precox the patient reveals himself by the reaction time (ordinarily not so evenly prolonged as in the depression of manic-depressive and, when not normal, more lawlessly desultory) and by the apathetic or perverse behavior toward the task set.

The association tests also help in differentiating the depressive states of manic-depressive insanity from some forms of *hysteria*. In the latter disease it is especially the emotional phenomena, the signs of the idea complexes emphasized by feeling (*gefühlbetonte Komplexe*), which are characteristic. In manic-depressive insanity the inhibition is demonstrable aside from the slowing effect upon the reaction time of such emotive complexes, and, further, the associations in general in this psychosis present a much less capricious and variable appearance. It must not be forgotten, however, that hysteria and manic-depressive insanity may be combined in the same patient.

**Anomalies in the Associative Connection of Ideas (Incoherence or Dissociation).**—Normally, the course followed in associations from the initial idea to the terminal idea is a well-regulated one, depending upon some dominant or superior idea. In many psychoses this normal connection of ideas in the associative processes is disturbed. The initial idea may be followed by a second idea which stands in no recognizable relation to it, an anomaly designated by psychiatrists as *dissociation* or *incoherence* of association. If a patient be asked what time it is, and she answer "fox," a severe disturbance of this sort is manifest. In milder cases the disturbance is manifested by the fact that the patient continuously loses the thread of thought. When a patient is disoriented as to time, place, and personal relations, along with general incoherence of association, the movements and speech of the patient being correspondingly disturbed (*motor incoherence*), the condition is technically known as *confusion* (*Verwirrtheit* of the Germans). Very often incoherence and confusion are used as synonymous terms.

It is customary to distinguish a *primary* from a *secondary incoherence*, the former appearing autochthonously, independent of any other psychopathic symptom (*e. g.*, in the so-called acute hallucinatory confusion or amentia), the latter as a sequel to other psychopathic phenomena (*e. g.*,

flight of ideas, hallucinations, strong emotions, dementias, etc.). The secondary dissociation or incoherence is, therefore, common to many severe mental disturbances. The process of loosening or dissociation has been designated *sejunction* by Wernicke. When a dissociation occurs suddenly and lasts only a short time, there result lacunæ in consciousness (the so-called *psychic eclipses* or *deliquia* of L. Meyer), which may form the starting point of manifold disturbances of the content of the ideas.

In mild instances of general incoherence a peculiar symptom is sometimes met with, known as *pathological notions* (*Einfälle* of the Germans). In the midst of a well-ordered and often extensive thought series, there comes suddenly and apparently without reason an idea or complex of ideas which stands in no connection whatever, by external or internal association as far as can be seen, with the former.

**Anomalies in the Intensity and Duration of Ideas.**—Instead of the ordinary course of associations, single ideas or idea complexes may attain to abnormal strength and be very frequently repeated in consciousness. Here belong the *exaggerated* or *hyperdynamic ideas* (*überwerthige Ideen* of Wernicke) and the *imperative ideas* (*Zwangvorstellungen* of Krafft-Ebing), sometimes known as *obsessions*. In individuals who are constitutionally psychopathic such ideas arise especially at times when they are mentally or physically exhausted. They are especially characteristic in the so-called *psychasthenic states* (Janet).

Wernicke distinguishes between hyperdynamic ideas, imperative ideas, and autochthonous ideas. The *exaggerated* or *hyperdynamic ideas* are characterized by the fact that the patients do not recognize them as intruders in consciousness; on the contrary, the patient regards them as the expression of his innermost nature, and in battling for them he is struggling for the maintenance of his own personality. They are, nevertheless, often felt as troublesome, and the patients frequently complain that they can think of nothing else. *Imperative ideas* or *obsessions*, however, are recognized as unjustifiable ideas, and are often spoken of as absurd. The *autochthonous ideas* have the attention forcibly directed toward them and are felt as troublesome intruders. Wernicke separates them from imperative ideas, since the latter are not regarded as foreign to the personality of the patient, and hence are not serious for the mental life of the patient as are autochthonous thoughts. The closest relations between hallucinations and autochthonous ideas have been pointed out by various psychiatric writers; indeed, transitions between the two sometimes may occur.

Many of the imperative ideas or obsessions are combined with marked affective disturbances, giving rise to the so-called *phobias*; thus, a patient may fear to cross an open place (*agoraphobia*), may fear shut-in places (*claustrophobia*), may fear contamination (*mysophobia*), may fear disease (*nasophobia*), or may be afraid of everything (*panphobia*).

When the same idea comes up over and over again the patients may be led to stereotyped methods of expression. The patient may answer a first question correctly, but may tend to give the same answer to various other questions subsequently put (perseveration).

In contrast with hyperdynamic ideas are the *hypodynamic ideas*. Certain experiences which in normal life may be well remembered and attended to are lost sight of or do not seem to have their normal value in the patient's mental life.

**Anomalies of Reproduction or Memory (Hypermnnesia; Hypomnesia; Amnesia; Paramnesia).**—The power of reproduction may be pathologically facilitated (*hypermnnesia*), diminished or retarded (*hypomnesia*), abolished (*amnesia*), or perverted (*paramnesia*).

*Hypermnnesia* is often met with in exalted or maniacal states and is usually associated with abnormal affective phenomena. Past acts, feelings, or ideas appear vividly in the mind, which in its natural state may have wholly lost the remembrance of them. Ordinary individuals sometimes experience hypermnnesia in sleep or in certain unnatural conditions, *e. g.*, under the influence of drugs (alcohol, morphine).

Weakened memory or *hypomnesia* may betray itself in the increased length of time it takes to recall an image or in the vagueness of the reproduction.

In outspoken pathological diminution or destruction of the power of memory (*amnesia*) there is complete inability for reproduction of a part or the whole of the past experiences. The term *amnesia* is by some reserved for complete loss of memory for a certain period; for example, that of dreamless sleep, of epileptic coma, of hysterical somnambulism, and the like. When the *amnesia* concerns not only the period of the pathological condition itself, but extends to a longer or shorter time preceding the attack, it is called *retrospective* or *retrograde amnesia*. The term *anterograde amnesia* has been used to designate the defect of memory which extends to occurrences of the period subsequent to the termination of the clouding of consciousness, a condition doubtless due to the depression of the recording faculty and the injury to the associative processes following the period of mental fog. When in the period subsequent to the termination of the mental fog the patient still remembers certain experiences, but soon after forgets them entirely, the condition is spoken of as *retarded amnesia*. Among the most interesting *amnesias* are those of the *alternating personalities* of hysterical patients, in which the first and third and second and fourth may stand in close associative connection with one another, while in the second and fourth states complete *amnesia* for the experiences of the first and third exists.

The *amnesia* may be partial, involving only the events of the recent past and not those of the distant past. Especially in progressive failure of memory, such as that met with in dementia, there is a gradual loss of memories, beginning with the most recent ones and involving gradually those of the past (*law of regression* of Ribot).

*Circumscribed amnesias* in special domains are met with very frequently in focal diseases of the brain. Thus, when optic memory images can no longer be called up, the condition is known as *mind blindness* (*Seelenblindheit* of the Germans). The mind-blind person may still have intact visual sensations, but he is unable to recognize what he sees because his visual memories are lost or, at any rate, cannot be awakened in the ordinary way by retinal stimuli, since the association between the primary

optic centres in the cortex and the areas concerned with visual memory are interfered with. Two great groups of cases can be distinguished from one another. In the first group we have to deal rather with a disturbance in optic perception than in optic memory. In this group belongs the so-called *apperceptive form of mind blindness* of Lissauer, which corresponds to the so-called *cortical blindness* (*Rindenblindheit* of Munk). In this group the principal disturbances are hemianopsia, diminution of visual acuity, and disturbance of color sense. Stereoscopic vision is also sometimes involved. On the psychic side there is usually an incapacity for orientation in space. There need not, however, be any marked disturbance of the visual memory or of the capacity for recognizing ordinary objects. Reading and writing may be nearly intact.

The second group of cases includes *mind blindness proper*, which is less a disturbance of perception than a loss of capacity for the intellectual valuation of retinal images. The condition is spoken of by Lissauer as the *associative form of mind blindness*. The patient is unable to recognize objects shown him because the visual images do not call up by association the memories which permit their identification, and so they appear to him as strange confusing figures (*optic asymbolia*). A patient looking at his wife, may know that she is a woman and see the details of her form and clothing exactly, but yet not recognize that she is his wife.

If very circumscribed areas in which optic memories are localized or the pathways to them be destroyed, the phenomena of word blindness or of optic aphasia may be met with. In *word blindness* or *alexia* there is an incapacity to read, although speech and the understanding of speech may be retained. The power to write may or may not be present. In the former case one speaks of *subcortical* or *pure alexia* (probably due to lesion of the the associative paths between the visual sense area in the cortex and the sensory speech areas). In the *optic aphasia* of Freund, a rare condition, objects held before the patient are seen and recognized but cannot be designated, although the patients can otherwise speak well and can find the particular word concerned when the stimulus is awakened through some other sense organ. It appears to follow most frequently upon lesions at the junction of the left occipital and left temporal lobes interrupting the association paths which extend from the two occipital lobes to the centre for word-clang memories.

A similar isolated loss of acoustic memorial images is known as *mind deafness*. The patient may hear everything, but cannot recognize the sounds. The most frequent form met with is that known as *word deafness* in the *sensory aphasia of Wernicke*. The patient hears the word, but no longer recognizes it and cannot understand it (lesion of the first temporal gyrus on the left side). Here, too, we probably have to distinguish a group of cases in which the disturbance is *perceptive* and a second group in which it is *associative*, the latter being sometimes referred to as *amnesic aphasia*.

When tactile memorial images can no longer be awakened we have *mind anesthesia for touch*. The patients feel contact, but cannot recognize objects through it. Similarly, there may be a loss of the memorial images for kinesthetic sensations.

Under the term *paramnesia* come the falsifications of memory, including what Sully has described as memory phantasms and memory illusions.

In the *memory phantasms* the patient seems to remember things which he has never experienced. The combinations of pure phantasy are regarded as actual experiences. They seem to be due to a loss of critical power, and depend upon enfeebled judgment. Here must be placed the tendency to confabulation and pseudo-reminiscences, so common in Korsakow's psychosis.

By *memory illusions* the memorial images themselves are falsified, the disturbance being due to faulty reproduction and lively fancy or the falsifying influence of a temporary affective state. As Sully puts it, the past now appears in the colors of the present. Such patients may appear to be liars, since they unconsciously distort their experiences in reproduction. A very good example of such illusions of memory is often seen in connection with the uncinate gyrus fits of H. Jackson.

A special form of memory deception is seen in the identification of a present situation with one presumably previously experienced. This *sentiment du déjà vu* is one of the most interesting paramnesias met with among psychasthenics. It seems to be as much a disturbance of perception as of memory. The patient, feeling that impressions escape him, comes to the belief that he is experiencing memories.

**Methods of Testing Memory.**—The tendency among neurologists at present is to treat all memory loss not as a disturbance of a unitary capacity of the brain, but rather as disturbances of a large group of single capacities. It is true that the memory as a whole, that is, the conglomeration of all these partial faculties, may be impaired particularly in some of the psychoses, but in neurological conditions in which circumscribed lesions of the cerebrum are met with we see more often injuries of the partial memory faculties. On testing the memory clinically, therefore, one must use methods which permit us to judge not only of the memory power in general but of the capacity for and fidelity of reproduction in each individual domain.

It is best to begin by testing the memory for fresh impressions or for recent events. One determines a defective power for the immediate acquisition and retention of ideas (recording faculty), and also defective capacity for the acquisition of general and relative ideas (power of abstraction). In addition, it is necessary to test the memory for older impressions or for events which have occurred earlier in the life of the individual, and here one does best to examine the patient's memory for what he has learned at school and to try his power of calculation. In testing school memories, one should have some knowledge of the extent to which the patient has been educated, as obviously the testing of illiterate persons for school memories would be useless. It is advantageous to use a uniform set of questions in order that comparisons of value may be made among the replies received from different patients. It is customary, therefore, to ask the patient to repeat a number of series which in school are learned by heart; for example, the alphabet, the numerals, the names of the months, and the names of the days of the week. Certain national and religious facts which everyone may

reasonably be expected to know may also be inquired into. To these questions may be added others regarding well-known facts of geography and history. Normal answers do not necessarily indicate mental normality since in non-paralytic dementias, for example, school memories are often very well retained.

The solution of the problems themselves is not the only important point. The time taken to solve them and concomitant phenomena of physiognomy and speech should be taken into account. The use of these methods has demonstrated that certain phenomena recur typically in different groups of clinical cases, and Sommer especially recommends the methods for the differential diagnosis of different forms of dementia. Through their application one obtains not only a more exact insight into the partial functions of the power of calculation, but also can give mathematical expression (where tests are repeated at intervals in the same cases) to the symptoms of periodic variation, of advancing intellectual disintegration, of stereotypism, etc.

**Falsification of the Content of Ideas (Delusions).**—The power of forming judgments and conclusions by means of the association of ideas depends in health upon the existence of certain regular relations between the various memories derived from preceding sensations regarding the body and the external world. Judgments depend, however, not entirely upon one's own personal experience, but upon tradition and upon the ideas dominant in the circle in which one lives at the time. One's own personal knowledge is supplemented by certain prevalent beliefs (religious, scientific, political, etc.). On account of the stronger feeling tone which accompanies beliefs their influence upon the power of judgment can scarcely be overestimated. While it is true that men of rich individual experience with strong powers of observation and analysis may attain to a high power of independent judgment; still no one, even the intellectually greatest, remains uninfluenced by belief, for individual experience, no matter how extensive, must be supplemented by beliefs which bridge over the gaps of experience. Everyone is, therefore, liable to make mistakes due to prejudice and superstition, and there is every transition from the mistaken judgments of human beings whom we call normal to the *delusions* or *false judgments* of the insane. In both instances the false judgment arises as a result of combinations of ideas which are out of accord with the facts of the so-called external world. The mistakes of the normal man can, however, be corrected by subsequent judgments depending upon new perceptions and more correct inferences, but the insane delusion is not amenable to such correction. Even in people who are not insane there may be certain false judgments which cannot be corrected. People brought up in an atmosphere of strong prejudice and superstition may be incorrigible in their beliefs and yet not be insane in the strict sense. Believers in Christian Science, in spiritualism, and the like, may form false judgments, but we would hesitate to call them insane. The false judgments are to be regarded as insane delusions when associated with other undoubted phenomena of alienation.

Insane delusions are divided into *primary delusions* and *secondary delusions*. The former usually arise from abnormal states of feeling in

connection with a pathological exaggeration of the consciousness of personality. The latter are, as a rule, explanation delusions or attempts on the part of the individual who has passed through a psychosis with injury to his mental processes to explain the changes in his consciousness.

In the primary delusions egocentric ideas dominate the consciousness and lead to the formation of false ideas of reference—that is to say, to false ideas regarding the individual and the processes of the external world. Such false ideas may, if there be a decided alteration in the associative mechanism, form the starting point for a permanently developing chronic delusional system (paranoid states). Clinically such individuals may appear to have normal recording faculty, recognition, and memory, but despite thereof manifest fundamental disturbance in the formation of judgments. On superficial examination such patients may seem very intelligent, and one would not suspect them of being insane until one learned from prolonged conversation the falsification of their judgments regarding their own personality and their entire *lack of disease insight* as regards such falsification of the content of their ideas. Temporary and curable delusions may occur in acute psychoses as a symptom of inhibition of normal associations, but the chronic, incurable delusional insanities depend rather upon permanent loss of certain possibilities of association, the patient being unable critically to sift and arrange his judgments as a normal individual can do.

Many of the chronic systematized and progressive delusional states begin with a period of psychic eclipse. The patients complain that for some days or weeks their minds have been empty and that they have felt restless. Some go so far as to assert that they must have been anesthetized or poisoned at the time. In the acute psychoses, especially those accompanying infections and intoxications, delusions are prone to arise as a result of the primary incoherence. The mixed-up ideas of the patient lead to peculiar and nonsensical judgment associations. In some cases delusions arise in which a primary associative disturbance cannot be demonstrated, the falsification of content of the ideas here depending upon pathological affective processes.

There can be no doubt that illusions and hallucinations are of great importance also for the origin of delusions. This is well illustrated in the so-called acute hallucinatory confusion or amentia; in curable cases the delusions disappear, but where a dementing process succeeds, the hallucinatory delusions may continue unaltered or develop farther.

In chronic paranoid states the delusions may precede the hallucination and illusions, the latter being the outcome of the former. In the so-called acute hallucinatory paranoia the delusions arise primarily (in contrast with amentia, in which they are more secondary); in this disease the primary suspiciousness of the patient may be followed by hallucinations which convert the suspicions into certainty.

Explanation delusions (secondary delusions in the strict sense) occur where pathological states of feeling are protracted (prolonged exaltation or prolonged depression) and form the basis of the false judgments.

The hyperdynamic ideas corresponding to an exalted state may easily lead to *delusions of grandeur*. Similarly, the hyperdynamic ideas cor-

responding to the depressed mood of the melancholic and the hypochondriac may become fixed as *depressive delusions*, which persist even after the pathological mood has disappeared.

The *expansive (megalomaniac) delusions* and *depressive (micromanic) delusions* together make up the *primordial deliria* of Griesinger. Depressive delusions include the delusions of sin, the hypochondriacal delusions, the poverty delusions, and the delusions of persecution.

*Delusions of persecution* differ from the other forms of micromanic delusions in that other people (enemies, persecutors) are held responsible therefor. Sometimes they arise primarily and are connected with abnormal sensations. The feeling of fatigue which sometimes follows the drinking of a glass of beer may excite the persecutory idea that the beer has been poisoned or that someone intended to drug the patient. Still more frequently persecutory ideas arise as a result of hallucinations or illusions. Complementary or contrasting persecutory delusions are often associated with delusions of grandeur, the patient assuming that he is being attacked on account of the greatness of his position.

*Ideas of reference (Beziehungswahn)* may be the forerunners of ideas of persecution. The patient refers looks, acts, remarks of other people to himself without sufficient reason. He thinks he is being influenced or watched or injured by someone else. He may become abnormally jealous (*jealousy delusion*) or he may get the idea that he is suspected of a crime and that he is to be brought into court (*imputation delusion*). The patient may attribute the persecution sometimes to definite persons about him, sometimes to invisible and unknown enemies. Sooner or later such a patient is likely to become convinced that a great conspiracy has been formed against him.

A word must be said about the so-called *general delusion of denial (délire de négation généralisé)* of the French. It usually follows delusions of sin, the patient becoming so bad in his own estimation that he identifies himself with Satan. He can atone for this only by eternal punishment, and so comes to regard himself as immortal. Such a patient may think that his body is infinite in size as well as in duration (*délire d'enormité*).

When delusions first appear they are usually accompanied by affective states which correspond to the content of the delusion, but after long standing this feeling tone of the delusions may disappear.

This mode of onset of a delusion may be sudden or it may develop gradually. Primary delusions tend to have the latter form of development, and are more apt to become fixed; hence the unfavorable prognosis in such cases, as a rule.

Fixed delusions are prone to be followed later by a so-called *systematization*, the patient adding complementary delusions to the fixed delusion, the new delusion being connected logically with the original false judgment. Such systematized delusional states may last throughout life.

To be distinguished from the delusions above described are the *defects in judgment* met with in imbeciles and patients, who for some reason or another have had some arrest of development. In such cases the memory pictures are defective and the associative relations among them are fewer than normal; as a result, incorrect judgments are very



likely to be formed. As Ziehen puts it, poverty in ideas and in associative connections among them, together with feebleness of judgment, are the essential features of the intellectual defect, both in *acquired* and *congenital feeble-mindedness*.

All grades of such feeble judgment are met with, from the mildest lack of critical power to the most complete lack of judgment. The former is due to absence of a few complicated ideas and associations, while the latter depends upon absence of the simplest everyday ideas and associations. Imperative ideas and delusions often disappear, but the feeble judgment once established nearly always remains.

**Anomalies of Affective and Emotional States.**—We have referred to the feeling tone accompanying sensations and ideas. Whereas sensations and their memory pictures (the so-called ideas or images) represent the elementary phenomena of consciousness which refer to objects external to a given perceiving subject or personality, the feelings, on the other hand, and the affective or emotional states in which they play the essential rôle, represent the more intimately subjective side of experience in that this portion of the content of consciousness expresses the attitude of the individual to objects, etc., in its most fundamental form; that is, in particular, as it bears upon his conduct.

The elements of our experience of external objects about which no difference of opinion exists are sensations. In the same sense *the elements of the affective side of consciousness* include *agreeableness* and *disagreeableness* (pleasure and pain of the older psychologists, and *positive* and *negative feeling tones* used by physiologists generally); these two at least, are elementary feelings. Whether Wundt's assumption that there are six directions in which elementary feelings may vary—the *agreeable* and *disagreeable*, the *exciting* and *depressing*, and the *straining* and *relaxing*—be correct or not need not here detain us.

*Emotions* are more complex than simple feelings, in that they contain not merely *affective*, but, quite as essentially, *sensational* (perceptual) elements also. The analysis of an emotion, for example, fear, reveals not only a feeling of disagreeableness, but also the sensational factors which represent the object which is feared, with complexes of kinesthetic and visceral sensations, the exact nature of which has not yet been discovered. The close relation between emotions and bodily movements is evident.

*Moods* are of the nature of emotions having the same general constitution, but, as a rule, they lack definiteness in that the sensational or objective side of a mood is not so frequently one particular object. Moods are, in general, of longer duration than emotions, and do not so readily express themselves in some definite act which tends to bring the state, as in emotion, to an end. On the contrary, they persist for longer periods as a kind of general affective tone of experience (*e. g.*, depression, exaltation). Manifestly, in both emotions and moods, memory and imagination are of, at least, quite as much importance as the sensations of the present moment, and in the majority of cases of much greater significance, since they permit of the constant development of the objective content of both emotions and moods entirely freed from objective control.

Feelings and emotions become associated with objects other than those originally experienced with them, and this association occurs at times in most fantastic ways. This fact of the *transfer of feelings* helps us to understand the origin and permanence of moods in both those of normal and abnormal mentality.

It has long been held that *the feelings have an important relation to the state of the body*; as far back as the seventeenth century one meets the view (in Hobbes) that pleasurable feelings are associated with bodily well-being, while disagreeable feelings are both expressive of and conducive to the ill-being of the body.

The effects of strong feeling and emotion upon the intellect are often marked, depending, of course, upon the intensity as well as upon the quality of the affective state. If not too strong, the association of ideas is favored and accelerated, but when violent there may be inhibition or even complete arrest. The positive affective states tend to increase the rapidity of associations, while those negative tend to retard them.

1. **Pathological Exaltation or Hyperthymia.**—This is best seen as a primary phenomenon in maniacal states. All the intellectual processes are accompanied by positive feeling tone. Even some of those which normally are associated with slight negative feeling tone appear in many to be accompanied by joyful feelings. It is not known whether the hyperthymia is really primary or is secondary to the psychic overexcitation. The experimental production of pleasurable feelings by the use of alcohol, opium, etc., supports the view that the affective state may be primary (Binswanger). Hyperthymic states may appear in the course of many psychoses, both acute and chronic. The hyperthymia is usually an episodic phenomenon. A form of secondary exaltation has been described in acute and chronic paranoid states. Here hallucinations and delusions of joyful content determine the affective anomaly (Ziehen).

2. **Pathological Sadness (Depression) or Dysthymia.**—This pathological predominance of negative feeling tones may be primary or secondary. In the *primary depression* the sadness may be out of all proportion to that which would normally accompany any sensations and ideas present. The depression may, indeed, be present without any apparent reason whatever. In addition to his general sadness the patient may present symptoms of *anxiety*. Usually this anxiety is accompanied by abnormal somatic sensations (precordial anxiety, epigastric pulsation, general unrest). Many have looked upon these somatic sensations as primary, but it seems more likely that they are cortical in origin.

The motor phenomena accompanying anxious states are characteristic. The rubbing of the hands, the restless movements of the legs, the rapid respiration, the palpitation of the heart, the constriction of the peripheral vessels are some of the motor and vasomotor phenomena accompanying anxious states. Such primary depressions with anxiety are usually followed by symptoms of mental retardation (thought inhibition) and the patient answers questions and performs the simplest calculations only very slowly. The general musculature of the body may be abnormally lax or abnormally rigid, as in the catatonic states. Where the anxiety movements of the body are pronounced, one speaks of *anxious*

*agitation*. Sometimes movement inhibition alternates with agitation of movements.

The patients are inclined to seek an explanation for their anxiety, and then arise the so-called explanation delusions. These may take the form of delusion of sin, of poverty, or of incurable disease. Primary depressions are met with as episodic states in various psychoses; in a mild form they are not infrequent in neurasthenia. The severest forms are met with in true melancholia. In the so-called *secondary depressions* the affective states seems to be directly dependent upon sensations and ideas with strong feeling tone.

**3. Pathological Irritability.**—The pathologically irritable person shows an abnormal tendency to anger and vexation. These affects appear too easily; exciting causes which in a normal person would not cause vexation call forth an affective reaction of abnormal intensity and long duration.

*Anger* is an affect which accompanies the tendency to attack persons or objects in the surroundings. Although it is a negative affect, it differs from sadness or depression in that the feeling of self is usually elevated. The association of ideas may at first be slowed, although as the anger grows there may be a sudden and explosive acceleration of the course of ideas. At the beginning there may be a tendency to motor inhibition, but after a time, as though through summation, actions are accelerated and are often explosively violent (raving mad).

During anger there is but little time for motives to come into play. The patient seems to fail to consider things at all; inhibitory ideas seem to have lost their power, or do not often come into his consciousness. His violence pays no regard to the welfare of his own person or of his surroundings. The energy of the movements may be greatly increased. The movements of anger are, however, so irregular as to be almost incoherent. This reveals itself in the speech of the angry man. He stammers and his sentences lack grammatical sequence or coherence (*anacoluthia*). It is not surprising, therefore, that after an attack of anger has passed individuals frequently show a partial amnesia of the motives leading to the passion, and even of their acts during the outbreak.

**4. Pathological Apathy, General and Circumscribed (Hypothymia; Athymia).**—In certain mental states feeling tones appear to be depressed or abolished. The condition is most pronounced in cases of melancholia. In many instances of neurasthenia the patients complain of loss of interest in things which normally should arouse them. Their feelings for their ideals have left them and they complain that to those dearest to them they have become indifferent. Some patients seem much exercised over this coldness of feeling; others show no depression on this account. An appearance of general apathy may be simulated when it does not exist. The non-participation of the patient in her usual interests may be due to a conscious suppression of emotional expressions, depending upon delusions, or it may be the result of a motor inhibition.

In contrast with this general apathy or depression of emotional excitability, as a whole, are the circumscribed defects in intellectual and reflected feeling tone met with especially in imbecility and dementia. In imbecility there is a faulty development of the emotional nature which

may keep pace with the defect in intellectual development or may exceed it, as in moral insanity. In imbecility of milder grade, anger, envy, hate, love, gratitude, hope, and fear may be well developed, but the individual does not attain to any intellectual interests. In spite of careful education it is impossible to arouse interest in art or science. In still milder cases the ethical or altruistic feelings alone may be deficient, the egoistic feelings being abnormally strong.

In acquired dementia, such as one sees in dementia paralytica, the loss of the higher feelings usually goes parallel with the loss of intellectual power. The changes in this disease show us very clearly the relation of conduct and behavior to our emotional life (altruistic, social, religious feelings). In alcoholic dementia, epileptic dementia, and in senility one meets with symptoms similar to those presented by the paralytic dement. In the later stages of these acquired dementias the whole emotional life may be undermined and a general apathy result.

5. **Pathological Instability of Feelings (Moods).**—In normal individuals, there is a certain persistence or inertia to emotional states. In mental disease the moods are often much more variable. Two forms of pathological instability of moods may be distinguished, a primary and a secondary. The *primary* form occurs most often in imbecility and dementia, where it is associated with intellectual defects. The loosening of the associative connection characteristic of the intellectual defect also accounts for the instability of the irradiated feeling tones. A new sensation or idea will then sometimes in a moment lead to new irradiations of feeling tones and cause a *bouleversement* of mood. This is best illustrated perhaps in dementia paralytica, in which a patient in tears or anger is by a suitably chosen word converted into a state of joyful exaltation.

In the *secondary* form of instability of moods the changeability of feeling is not a primary affective disturbance, but is secondary to a pathological inconstancy and incoherence of the contents of the ideas or sensations of the patient. Where the ideas are constantly changing the moods also change. This is well illustrated in hysteria and in some forms of paranoid states. A pathological capriciousness is very characteristic of many hysterical patients.

6. **Pathological General Increase of Affective Excitability.**—In contrast with general apathy, a general increase of affective excitability seems sometimes to be met with. The individual is abnormally susceptible for all emotional impressions. This is the case in some instances of neurasthenia, but it is also occasionally met with at the beginning of severe organic psychoses. When the increase in excitability is limited to the so-called higher feelings—ethical, esthetic, and religious—it is designated as *pathological transport* or *ecstasy* (*krankhafte Ergriffenheit*). Such patients become over-enthusiastic for political, religious, or humane movements. When delusions develop upon the basis of this pathological ecstasy an *eknoic state* is said to exist.

The so-called *imperative affects* are those pathological feelings which arise without cause, and which the patients themselves designate as strange or compulsory and independent of imperative ideas, delusions, or hallucinations. A patient in good humor may suddenly and without

reason have a strong sympathy or a strong antipathy for some person near him, combined with a feeling that some force has artificially brought about such a tendency in him.

**Anomalies of Conduct (So-called Will or Volition).**—Psychologists no longer recognize will as a special “faculty” of the mind. Conduct is, for them, the direct result of the total content of consciousness (sensations, percepts, memories, imaginations, feelings, emotions) of the moment. In states in which there is a general increase of motor action in mental disease we speak of *motor agitation*; where motor action is diminished or abolished, as a whole, we speak of *aboulia* or *motor stupor*.

In observing anomalies of volition one should notice not only the ordinary voluntary acts, but also alterations in speech, in expression, and in gesticulation. The so-called voluntary movements depend directly upon an associative process leading to some goal idea, while the expressive movements (mimic and pantomimic) appear to be determined chiefly by the intensity and quality of the feelings. Disturbances of sensation, of memorial reproduction, affective disturbances, as well as of the association of ideas proper, can all influence conduct. Of the sensory disturbances which influence volition, hallucinations and illusions are most important. A sudden hallucination may lead to a wholly unexpected act of violence.

Wernicke has classified movements (outside of the reflex movements) into expressive, reactive, and initiative movements. By *expressive movements* are meant all those by which the affective and emotional states of a person are manifested. The *reactive movements* are those which follow direct external stimulation (answer to questions by speech or other movement; behavior on physical examination). The *initiative movements* are those which arise apparently spontaneously and not as a result of an immediate external stimulus, the whole conduct, behavior, action of a person in a certain situation. All movements not reflex, expressive, or reactive belong to the initiative group. Wernicke divides all psychomotor disturbances into (1) those in which there is lowered excitability or conductivity of nerve paths (*akinesis*); (2) those in which there is increased excitability or power for conduction (*hyperkinesis*); and (3) those in which there is a perverse excitability (*parakinesis*).

Action may be pathological on account of the absence of ideas which are present in normal states; this absence of ideas may be due to defective development (imbecility) or to loss of memory images, following upon disease (acquired dementias). Pathological actions of this sort are designated *defective acts*. They often resemble the normal acts of lower animals. They may be sly and skilful, but are pathological in human beings on account of the fact that they are uninfluenced by higher and more abstract ideas.

In the motor agitation of *mania* the pleasurable feelings lead to remarkable mimic and pantomimic motor discharges and to a logorrhœa characterized by rhymes and alliterations.

In the milder forms of maniacal exaltation in which the associative connection of the ideas is still fairly well retained, initiative movements occur more rapidly and in greater numbers than normally, and lead to

conduct which, although apparently consistent, is absurd in its motives and aims. In the severer outbreaks, stormy, impulsive acts result from the hodge-podge of ideas which swim through the consciousness.

The inhibiting and slowing of motor action which accompanies negative feeling tones is characteristic of *melancholic states*. In simple depression pantomimic movements (gesticulation) are reduced to a minimum. The voluntary muscles are generally relaxed, the arms hanging loose by the sides or the hands folded on the lap. The head sinks upon the breast, the eyes are lowered, and there is a tendency to convergence. The mandible falls and the angles of the mouth are drawn downward. The lid slits are narrowed and the eyebrows lowered except at their medial extremities. Usually the patients do not weep; indeed, they sometimes complain of an inability to weep. Often the secretion of the lacrimal glands is actually diminished, accounting for the lack of lustre of the eyes. Depressed patients have little to say, and speak in low tones.

The influence of depression upon motor activity becomes changed as soon as a state of *anxiety* is added to the depression. At first anxiety increases the inhibition of cortical associations and tends also to slow action, but as the anxiety increases there arises the idea of flight, and the patient seeks in every way deliverance from his anxiety. He cannot rest and is impelled to walk about day and night, lamenting his state (*motor agitation of anxiety*). It is here that the patient so often seeks relief in *suicide*. Perfectly quiet for a time as a result of motor inhibition, the anxious patient may suddenly make a suicidal attempt, or become violent. Other patients try to still their anxiety by alcoholic excess or onanism. These attacks due to anxiety occur in paroxysms. Between the paroxysms the motor inhibition of depression is resumed.

The behavior of apathetic patients varies according to the extent of the apathy (general or partial). In *general apathy* motor activity is reduced to a minimum. Since, normally, acts occur only when movement ideas with positive feeling tones arise in consciousness, acts cease in general apathy because most of the movement ideas which arise in consciousness are devoid of feeling tone (*apathetic motor stupor*). In *partial apathy*, where certain only of the feeling tones are lacking, it is the conduct which depends upon the presence of more complex and especially the ethical ideas which is most prone to be defective. The patient acts wholly according to his lower egoistic interests, not distinguishing between right and wrong.

In general apathy all the muscles of the body are lax; even the cheeks hang down. The head and limbs assume positions which depend upon gravity. The upper lids fall so as to simulate ptosis. The patients incline to lie down much of the time; in severe forms the apathy may resemble sleep. The condition of the pupils distinguishes the pseudo-sleep of apathy from true sleep; in the latter, when the eyelid is opened there is at first dilatation and subsequent contraction, while in apathy the pupils are usually midway between dilatation and contraction and there is a distinct contraction when light enters the eye on opening the lid.

**Acceleration of Motor Actions**—When motor discharges occur more rapidly than normal we speak of *motor agitation*, or sometimes of patho-

logical *pressure of activity*. One meets with every degree of it from abnormal talkativeness with overactive facial expression to the enormous acceleration of speech known as logorrhœa. The mimic and pantomimic movements may be so much exaggerated that we have excessive grimacing and violent gesticulation. The patients are restless, they assert that they cannot sit still, and they spend their time pacing up and down the room or wandering about. Such patients may be pathologically busy, beginning a dozen and one tasks hastily and enthusiastically but quickly turning from each.

In the severest forms of motor agitation when the patient becomes "raving mad," his cries may become inarticulate and his movements violent and purposeless. Such a patient may tear up his clothing or his bedding, break the furniture of his room, and attack people near him.

Psychiatrists distinguish a primary from a secondary pressure of activity, the former corresponding to primary flight of ideas and occurring with it, the latter depending upon hallucinations or violent affects (*hallucinatory agitation* and *affective agitation*).

**Retardation of Motor Actions.**—Slowing of motor acts accompanies depressed states, which are accompanied by thought inhibition. When there is motor inhibition, together with thought inhibition and aprosexia, the condition of stupor is said to exist. One meets with mild and severe grades of *motor stupor*. In the milder forms movements seem difficult and are carried out more slowly than normal. The patient understands questions or commands less promptly than when he is in health. He pronounces the words of the answer more slowly, and any movements he makes are more deliberate than they should be. Mimic and pantomimic movements may be suppressed entirely. In his every-day life the patient manifests feeble will (*aboulia*), and is the victim of indecision.

When the motor stupor reaches a high grade it may assume any one of three different forms:

1. There may be complete relaxation (*resolution*) of the body musculature, the patient lying for weeks or months without stirring a limb, with complete absence of resistance to passive movements, the eyes kept closed.

2. There may be a general rigidity or tension of the muscles of the body (*catatonic rigidity* or *attonnity*), the body being held stiffly in a general position of flexion (flexion type) or of extension (extension type), or in various bizarre attitudes, the eyes held tight shut, the teeth clinched. Passive movements are strongly resisted, and, as a rule, attempts at passive movement increase the general rigidity. The patients are mute and in general *negativistic*. Sometimes, however, there is a peculiar susceptibility to motor suggestion known as *command automatism*. On passive movement the limbs move like wax, retaining the position in which they are put often for three or four hours (*flexibilitas cerea*); sometimes they repeat movements made before them (*echopraxia*), or repeat words pronounced in their hearing (*echolalia*).

3. There may be a limitation of initiative movements to a few acts repeated continuously for hours, days, or months at a time (*stereotyped*

*movements*), the patient rocking to and fro or from side to side, or continuously rotating the head or the trunk.

One tries to make out in each case whether the motor inhibition is primary (*true stupor*) or secondary (*pseudo-stupor*). *Primary motor stupor* corresponds to primary depression of thought activity, being part of the general slowing of associative processes (*e. g.*, in melancholia). Usually it takes the type of relaxation of the muscles or of simple catatonic rigidity. In *secondary motor inhibition* the pseudo-stupor may depend upon definite hallucinations, delusions, or affects.<sup>1</sup>

**Incoherence of Motor Actions.**—When there is incoherence of the association of ideas the motor acts also become incoherent (*motor incoherence*); the patient is in a state of confusion. In its mild form it betrays itself in a peculiar lack of plan in the more complex acts of the patient. On going for a walk he may wander hither and thither without aim and out of accord with the purpose which he at first had in mind. Such a patient will busy himself first with one thing and then with another in a most unsystematic way.

In the worst forms of confusion the acts become nonsensical and stand in no correct relation to the ideas preceding them (*parapraxia* and *apraxia*).<sup>2</sup> The patient does not know how to use simple objects. He cannot light a match or open a knife, may try to eat milk with a fork, and may bite his own finger instead of a morsel of food held in his hand. The confusion may extend to speech and writing, the patient designating objects falsely (*paraphasia*, *paragraphia*). The movements of the body may become incoördinated, resembling ataxia or chorea. Even the mimic movements may no longer correspond to the underlying affect. The patient's grimaces are out of accord with his feelings; his laughing and crying stand in no ascertainable relation to the content and feeling tone of his ideas (*paramimia*).

Motor incoherence is sometimes combined with motor agitation, the mixed state being designated *incoherent agitation* and the excessive incoherent movements described as *jactitations* (chorea magna of the older writers). When this is accompanied by fever the condition has been called *delirium acutum*.

The motor incoherence is called primary when it cannot be attributed to other psychopathic states (*e. g.*, in the incoherent form of paranoia). It is called secondary when it is due to hallucinations, delusions, flight of ideas, affective disturbances, or imbecility.

**Anomalies in the Intensity and Duration of Acts (Change of Acts).**—In health initiative movements undergo changes corresponding to the changes which take place in the sensations and ideas in consciousness. When special motives are present certain movements may become dominant. In mental disease, however, dominant movements or acts may appear without adequate motivation (*hyperdynamic acts*). These hyperdynamic acts may be *primary* (not due to other psychopathic symptoms). A good example is seen in the so-called ties. Most hyper-

<sup>1</sup> For the differential diagnosis of the different forms of stupor the reader is referred to Ziehen's *Psychiatrie*, second edition, Leipzig, 1902, p. 156.

<sup>2</sup> Wilson (S. A. K.), *Studies in Apraxia, Brain*, 1908.



dynamic acts are, however, *secondary* to delusions, hallucinations, or anxious states. The monotonous recurrence of a certain act or attitude (*motor stereotypy*) is probably due to a more or less permanent psychomotor hyperdynamy.

Among the interesting stereotyped movements frequently met with are the snout-like movements of the lips (*snout cramp*) and constant blepharospasm. Sometimes the stereotypy manifests itself in certain bizarre movements, the so-called *mannerisms*. Many patients have a stereotyped scanning speech or vary the pitch of their voice in monotonous repetition. Sometimes single syllables or sounds are intercalated in the speech, a habit usually regarded as affectation, but spoken of by psychiatrists as *stereotyped embolophrasia*.

When a motor innervation has been completed and there is a tendency on the part of a patient to repeat it afterward, even when other movements are required of him, the condition is designated *motor perseveration*. A common example is the repeated showing of the tongue, after one has asked to see it, even when the patient is subsequently requested to show his teeth, to beckon with his finger, or to close his eyes. This is to be distinguished from the stereotypy which has its origin within.

Temporary motor hyperdynamy reveals itself in the so-called *impulsive acts*, due to a sudden affect, delusion, imperative idea, or hallucination.

**Anomalies of Conduct Due to Delusions and Imperative Ideas or to Imbecility and Dementia.**—A patient who has delusions or imperative ideas will often reveal the character of the delusions or the ideas in his conduct. The whole facies and the attitude of the patient with delusions of grandeur are characteristic and are in marked contrast with the appearance and behavior of the patient suffering from delusions of self-depreciation. When delusions of persecution exist, the patient is ever on the defence (bolting of doors, sudden flights to escape enemies). Sometimes the *persecuté* becomes the *persecuteur*; then, instead of defensive movements, he may assume the attitude of attack. Such patients are usually dangerous in the community (paranoia).

The so-called *imperative acts* are the result of the imperative ideas also previously described. Here the patient recognizes the absurdity of his act, but performs it all the same. One should distinguish carefully the imperative idea which is accompanied by the impulse to motor action and that not associated with such motor impulse. In the former case the imperative act frequently results, in the latter it does not. In mysophobia the continuous washing of the hands for hours at a time is an illustration of an imperative act. A child's imitation of its parents may be regarded at a certain stage of development as a normal echokinesis, but later on direct imitation of movements observed ceases to be desirable, and the tendency is normally suppressed.

The influence of defective judgment upon the conduct becomes obvious in the study of imbecility and the various forms of dementia. The acts of defectives reveal the absence of those complex ideas which normally determine the play of our motives. There is an absence of due consideration before action. The so-called criminal acts doubtless belong here.

**Disturbances of Motility Proper.**—In the preceding sections motor disturbances of psychic origin have been referred to. We have now to consider the disturbances of motility less directly psychic or entirely infrapsychic in their origin, disturbances due to alterations in function of the motor pathways from the cerebral cortex to the muscles, or to alterations in the cortex itself or in the muscles themselves.

**Atrophy and Hypertrophy of Muscles.**—The *state of nutrition of the muscles* yields important clues to the nature of many nervous diseases. *Muscular atrophy* may or may not be associated with paralysis of muscles. *Simple atrophy* of the muscles should be distinguished from so-called *degenerative atrophy*. In the former there is only a quantitative diminution in size; in the latter there is in addition an actual degeneration of the muscle substance. Degenerative atrophy occurs in lesions of the lower motor neurones (anterior horn cells, motor nuclei, or cerebral nerves, peripheral motor nerves). Simple atrophy may be due to disease of the muscles themselves or to lesions of the motor conduction paths situated above the lower motor neurones (*e. g.*, pyramidal tract lesions). It may be due simply to disuse. In true degenerative atrophy the so-called reaction of degeneration is found. Where the degenerative atrophy is due to lesion of the anterior horns or of the motor nuclei of the cerebral nerves, *fibrillary twitching* is commonly present. It is rare to find it in the lesions of the peripheral motor nerves.

The muscles may be the seat of a true *hypertrophy* in athletes. There is localized hypertrophy of muscles which are subject to cramp-like processes which continue a long time. A condition resembling hypertrophy of the muscles, but which in reality is due to increase of connective tissue and fat, is seen in certain types of muscular dystrophy (so-called *pseudo-hypertrophy*).

**Musculature Tone, Atony, Hypotony, Hypertony, Contractures.**—The condition of *tone (tonus)* presented by the voluntary muscles is of importance. When the resistance to passive movement is abnormally small, we speak of *hypotony* or *atony* or *resolution* of the voluntary muscles. When there is no abnormally strong resistance to passive movement, we speak of rigidity or *hypertony* of the muscles. It is conspicuously present in the various spastic paralyses, in many motor spasms, in catatonia, etc. When along with hypertony a muscle remains permanently contracted, we speak of *spastic contracture*. When certain muscles are paralyzed and on subsequent voluntary innervation only the antagonists contract, there comes a time when relaxation of these antagonists no longer results in a normal position of the limbs. Such a permanent shortening of the antagonists is sometimes spoken of as *paralytic contracture*. In contrast with these *neuropathic contractures* (spastic and paralytic) there occur also *myopathic contractures* due to inflammation of the muscles and tendons, *contractures from shortening of fascia* as a result of scars, *contractures of reflex origin*, and *hysterical contractures*.

**Paralyses.**—Inability to bring the muscles to contraction is known as paralysis. Paralyses may be *classified* in different ways: (*a*) According to the state of nutrition of the muscles; (*b*) according to the state of tonus of the paralyzed muscles; (*c*) according to the distribution of the paralysis.

As regards the classification according to the *state of nutrition of the muscles*, paralyzes may be divided into (1) atrophic, and (2) non-atrophic paralyzes. By *atrophic paralysis* is meant a special form not due simply to disuse and accompanied by rapid reaction of degeneration. It indicates a lesion of the lower motor neurones, either in their nuclei of origin or in the peripheral nerves. By *non-atrophic paralysis* is meant the form in which the paralyzed muscles undergo but little shrinking in volume; no reaction of degeneration can be made out.

According to the *state of tonus* of the paralyzed muscles, paralysis may be divided into (1) flaccid paralyzes, and (2) spastic paralyzes. Flaccid paralyzes are usually associated with loss of the deep reflexes, while spastic paralyzes are usually associated with exaggeration of the deep reflexes. The flaccid paralyzes are due, as a rule (although not always), to lesions of the lower motor neurones. The spastic paralyzes are due nearly always to lesions of the upper motor neurones. As regards the *distribution of the muscles paralyzed*, paralyzes are divisible into: (1) Neural paralyzes; (2) plexus paralyzes; (3) radicular paralyzes; (4) paralyzes of one extremity or of the face (*monoplegias*); (5) paralyzes of half the body (*hemiplegias*); (6) paralyzes of both lower or both upper extremities (*paraplegias*).

In the *neural paralyzes* a single muscle or the muscle innervated by a single nerve may be involved (*e. g.*, Bell's palsy). In the *plexus paralyzes* the whole or a part of the muscles supplied by the brachial or lumbosacral plexuses may be involved. The paralyzed muscles in *radicular paralyzes* correspond to the motor innervation from single nerve roots. The *monoplegias* may be due to lesions of peripheral nerves or of the central nervous system. The face may be alone affected (*monoplegia facialis*); if it be the arm alone we speak of *monoplegia brachialis*; if the leg alone, of *monoplegia cruralis*. When the musculature of one-half of the body is paralyzed the condition is known as *hemiplegia*. If the muscles are only weakened, the term *hemiparesis* is employed. A hemiplegia may be organic or functional; it may be flaccid at first but is usually spastic later; it may or may not be accompanied by sensory disturbances (*hemianesthesia*; *hemianopsia*). It may be cortical, capsular, peduncular, pontile, or medullary in origin. A hemiplegia may be associated with paralysis of the muscles supplied by the opposite oculomotor nerves (*hemiplegia alternans superior*, or *Weber-Gubler type*); or the face may be paralyzed on one side and the arm and leg on the other (*hemiplegia alternans inferior*, or *Millard-Gubler type*).

The term *paraplegia* is usually restricted to paralysis of both lower limbs, with more or less involvement of the trunk muscles. The paraplegia may or may not be associated with sensory changes; it may be organic or functional, flaccid or spastic. The flaccid paraplegias may be due to disease of the spinal cord or the peripheral nerves, while the spastic paraplegia is due to lesions of the upper motor neurones, either in the spinal cord or at a higher level. The Brown-Séquard syndrome is sometimes spoken of as a *hemiparaplegia*. When all four extremities are paralyzed, owing to a lesion in the cervical cord, the paralysis is spoken of as a *cervical paraplegia*.

**Motor Irritation.**—The various forms met with include: (1) Tremor. (2) Fibrillary twitchings. (3) Athetoid and choreiform movements. (4) Clonic and tonic spasms.

By *tremor* is meant rhythmical, involuntary, frequent oscillatory movements of slight extent. The tremor may be fine or coarse; thus, that of Graves' disease is very fine and the rate varies from 8 to 10 oscillations per second; in paralysis agitans, on the other hand, the tremor is coarser and the oscillations less frequent (2 to 4 per second). A tremor which appears only when voluntary movements are undertaken is known as an *intention tremor*. Closely related to it is *nystagmus*.

By *fibrillary twitchings* are meant the contractions of single fibre bundles of any given muscle, the contractions being insufficient to lead to any locomotor result. Fibrillary twitching is a totally different thing from tremor. It is met with especially in disease processes in which the muscle is degenerating, owing to lesion of the cell bodies of the lower motor neurones.

By *athetoid movements* are meant certain slow, involuntary movements of extension, flexion, adduction, and abduction, each individual part moving by itself independent of the others, so that the various parts may at any given moment occupy very different relative positions in space. The movements may be unilateral (*hemiatheosis*) or bilateral. Such movements are prone to occur in lesions of the corpus striatum. They do not seem to be prominent, however, in the progressive degeneration of the lenticular nucleus described by S. A. K. Wilson.

*Choreiform movements* include the involuntary, quickly changing movements, made without plan or purpose, met with in various diseased conditions. They may involve any of the voluntary muscles of the body. This fact, together with the greater quickness of the movements, distinguishes them from athetoid movements. The movements are best studied in *chorea minor* (St. Vitus' dance; Sydenham's chorea). A unilateral chorea is not infrequent after hemiplegia (*post-hemiplegic hemichorea*). In hysteria a very coarse form of chorea known as *chorea major* is sometimes met with. It is frequently associated with *clownism*; in the same disease a form of chorea occurs in which there are quick, lightning-like contractions in single muscles similar to those which follow upon electrical stimulation (*chorea electrica*).

Involuntary movements of certain muscles which accompany voluntary movements of other muscles are known as *associated movements* (*muscle synergies*). One sees such movements frequently in the paralyzed limb of hemiplegics on strong voluntary innervation of the healthy limb. The *tibial phenomenon of Strümpell* may be regarded as an instance of a spinal associated movement due to injury of the pyramidal tracts. When it is present flexion of the lower extremity at the hip-joint and knee-joint calls forth a dorsal flexion of the foot and elevation of the medial margin of the foot, despite the patient's effort to suppress these movements.

*Spasms and convulsive movements* are among the commonest instances of motor irritation. By *clonic spasms* are meant involuntary quick jerks of the muscle which follow one another rapidly without interruption.

If the distribution includes a large number of muscles at any one time, we speak of *clonic convulsions*. In *tonic spasm* the individual muscular contractions last a long time. When a single muscle is involved with severe pain the condition is known as *cramp*. When muscle groups are affected together, or the whole body is involved, we speak of *tetanus* and *tetanic contractions*.

*Convulsive seizures* or *convulsions* occur in epilepsy, hysteria, tetanus, and eclampsia. In the ordinary *epileptic attack* the convulsion consists at first of a tonic spasm of the whole body musculature, followed after a short time by clonic spasms. In cortical irritation a peculiar type of epileptiform convulsions known as the *Jacksonian attack* occurs. The muscular contractions begin in single muscle groups and radiate into other motor domains (so-called "march" of the convulsion). In hysteria the convulsive movements may resemble those of epilepsy, although they are usually much more violent and the movements more varied.

In *tetany* we meet with intermittent tonic spasms of bilaterally symmetrical groups of muscles, associated with painful sensations and paresthesias. The attitude of the hands and forearms is especially characteristic ("*obstetrical hand*"), but muscles in various parts of the body may be attacked. Even when the spasm has passed off in the arms it may be produced by applying pressure to the arm above the elbow with a blood pressure apparatus (*Trousseau's phenomenon*). The mechanical excitability of the muscles of the face is greatly increased. Mere stroking of the cheek or tapping upon the branches of the facial nerves (*pēs anserinā*) with a percussion hammer calls forth quick muscular contractions in the face (*Chvostek's phenomenon*). The sensory nerves may also be hypersensitive, and tapping at Valleix's points then calls forth abnormally intense sensations (*Hoffmann's symptom*). On electrical stimulation the motor nerves are found to be hyperexcitable in tetany (*Erb's phenomenon*).

In Thomsen's disease (*myotonia congenita*) there exists a so-called *intention rigidity* of the muscles. On voluntary contraction of any group of muscles there results an abnormally strong contraction of long duration which leads to motor inhibition. Gradually the muscles relax again.

In psychasthenia and in certain other psychoneurotic states sudden contractions of the muscles known as *tics* are met with. In the so-called *maladie des tics* of Gilles de la Tourette the musculature of the whole body may be involved, and the condition is associated with mental deterioration.

**Disturbances of Coördination (Ataxia).**—The mechanism by which muscles act together for purposeful effects is known as *coördination*. Almost every voluntary movement requires the simultaneous and successive activity of several muscles (*synergists*); some of the muscles are contracting (*agonists*); others are relaxing (*antagonists*). The grouping of contractions, their succession, and the force of each have to be carefully regulated in order that the movements shall be harmonious and purposeful. Disturbance of this mechanism leads to the anomaly of movement known as *ataxia*.

The presence of ataxia is not necessarily combined with weakness of the muscles. It is, however, frequently associated with loss of deep sensi-

bility and with hypotony of the muscles. It seems certain that ataxias are due more to interference with the centripetal or sensory paths than to interference with motor conduction paths.

In the so-called *cerebellar ataxia* there is a disturbance of equilibrium, manifested especially on standing and walking. The patient stands with his feet wide apart and sways on walking from side to side like a drunken man. His tendency on walking is to let his legs run ahead of his body (*asynergie cerebelleuse* of Babinski). In trying to rise from the recumbent position, the patient, instead of lifting his trunk, is likely to lift his legs in the air.

In cerebellar disease, the patient lying on his back with the lower extremities in the air, the thighs flexed and the legs abducted, can maintain sometimes a *fixation of position* beyond that possible for a normal man; the lower extremities behave as though cataleptic (Babinski). On the other hand, the capacity (*diadokokinesis*) to carry on quickly a series of antagonistic movements, such as rapidly alternating pronation and supination, may be lessened (so-called *adiadokokinesis*).

**Electrical Condition of the Muscles and Nerves.**—Much information of value concerning the state of the muscles and motor nerves can be arrived at by careful electrical examination. One determines the *excitability and conductivity of the motor nerves* and the *direct excitability of the muscles*.

*Increased excitability* to electrical stimulation is not very common. It is occasionally met with in a beginning neuritis, in tetany, in beginning dementia paralytica, etc. A *lessened excitability* is met with in all old paralyses due to lesions of the upper motor neurones and also in the myopathic form of muscular atrophy.

The most important electrical reaction for diagnostic purposes is the so-called *reaction of degeneration* (*De R*). When it is complete, changes in the electrical excitability, both in the nerves and muscles, are demonstrable. The excitability of the nerves for both faradic and galvanic current grows less and less until finally it disappears. The excitability of the muscle decreases for the faradic current *pari passu* with that of the nerves until it also is finally abolished, but the excitability to the galvanic current undergoes a remarkable change. Instead of a quick, lightning-like contraction the reaction to the galvanic current becomes slow, worm-like, and long drawn out, and the current, when applied directly to the muscle, shows an increased excitability of the latter, very feeble currents calling forth contractions. In addition there is a *reversal of the normal law of contraction*; normally, the cathodal closure contraction can be produced with the feebler current, whereas in reaction of degeneration anodal closure contraction is more easily produced than cathodal closure contraction, and the cathodal opening contraction approaches in case of producibility that of anodal opening contraction.

When the reaction of degeneration is present it is proof positive of a lesion of the lower motor neurones, although it may affect either the cell bodies of the neurones, the peripheral motor nerves, or the nerve endings in the muscles; thus, a reaction of degeneration is common in anterior poliomyelitis, in progressive muscular atrophy, in syringo-

myelia involving the anterior horns, in the various forms of neuritis, and in diseases affecting the bulbar motor nuclei. There is no reaction of degeneration in lesions of the brain and cord which do not involve the lower motor neurones, nor is it present in the so-called pure primary muscular atrophies (dystrophies).

In myotonia congenita the so-called *myotonic reaction* of Erb is obtained which is peculiar in this, that the muscular contraction lasts for a long time on direct faradic and galvanic stimulation after the current is again opened, and, besides, peculiar wave-like contractions can be produced if one stimulates the muscle near its insertion, placing the other electrode near the origin of the muscle.

In myasthenia gravis the so-called *myasthenic reaction* of Jolly can usually be obtained. A tetanizing faradic current when first applied shows the presence of normal excitability, but gradually the response diminishes and in a short time no response can be elicited.

**Anomalies of Gait.**—The limits of normal variation in gait are wide and several tolerably characteristic gaits are to be distinguished. Among the more important are the wobbly gait, the paretic gait, the spastic-paretic gait, the spinal ataxic gait, the cerebellar ataxic gait, the gait of tremor, the hemiplegic gait, and the gait of intermittent claudication.

The *wobbly gait* is often due to paralysis or atrophy of the *M. gluteus medius et minimus*. It is also seen in congenital dislocation of the hip-joint.

The *paretic gait* assumes two types, the simple paretic gait and the partial paretic gait. In the *simple paretic gait*, due to muscular weakness only, the movements of walking are slowed and the steps are shortened. Frequently there is an exaggerated flexion at the knee-joints. In severer cases the patient has to walk with crutches. In the *partial paretic gait* certain only of the muscles are weak. The most common form is that due to paralysis of the peroneal muscles on both sides. There is toe-drop, leading to lengthening of the leg; in order to compensate for this the lower extremity must be overflexed at the hip and knee. This gait was described by Charcot under the name of *steppage*.

In the *spastic-paretic gait* the weak muscles are hypertonic and the stiffness slows the movements and diminishes the excursions. The lower extremity moves more or less, as a whole. The toes cling to the ground; the difficulty in flexing the knee and hip is partly overcome by elevation of the pelvis on the side of the swinging leg. Often there is adductor spasm in the thighs, so that the knees rub against one another, and there is a tendency of the legs to cross on walking. In hemiplegia with spasticity the rigid limb swings lateralward, making a movement of circumduction, distinguishing it from the gait of hysterical hemiplegia, where the paralyzed leg is "dragged" forward. In cerebral softening and especially in pseudo-bulbar paralysis the steps are often very short, the foot being lifted from the ground only with difficulty. This gait is spoken of as *demarche à petits pas*.

The *spinal ataxic gait* is very characteristic and is easily recognized. The excursions of the movements are all exaggerated, the hip is overflexed, and rotated lateralward, the toes are lifted and the whole leg suddenly thrown forward, the foot being brought to the ground with a

stamping sound. The feet are kept wide apart and the patient watches his movements closely, being almost sure to fall if he looks away.

Two kinds of *cerebellar ataxic gait* are described, one due to disturbance of equilibrium and often associated with vertigo, the other depending upon a movement ataxia. In the former, the gait resembles that of a drunken man, the patient swaying in a very irregular manner. In the latter, the patient walks with his feet far apart, stamping on the ground, but without the wide excursions of the spinal ataxic gait.

The character of the *gait in tremor* depends upon the cause of the tremor. Gaits more or less characteristic are met with in multiple sclerosis, in hysteria, and in paralysis agitans. In the latter, in addition to the stooped attitude of the patient, we meet with the phenomena of *propulsion* and *retropulsion*; the patient walking forward or backward has difficulty in stopping himself. In hysteria inability to stand and walk is sometimes a prominent symptom (*astasia-abasia*).

**Anomalies of Speech and Writing.**<sup>1</sup>—Disturbance in the articulation of the speech sounds is known as *dysarthria*. In its highest grade speech sounds can no longer be emitted (*anarthria*). Dysarthria may depend upon weakness of the muscles of the lips, of the tongue, of the velum palatinum, or of the larynx. The speech has a *nasal twang* in cleft palate and in paralysis of the velum palatinum. It is often heard in patients with adenoids. In bulbar paralysis it is the *lingual* letters (s, l, d, t, n) which are difficult at first; later the *labials* (p, b, m, f, w, o, u) are affected, and finally the *gutturals* (g, k, ch, r) become indistinct.

When the speech is especially slowed (convalescence from acute disease, psycho-motor retardation) the condition is known as *bradylalia*. When the individual syllables are separated by abnormally long pauses, the condition known as *scanning speech* is said to exist. It is especially characteristic of multiple sclerosis.

In *stuttering* there are spasmodic contractions of some of the speech muscles, interfering with production of the speech sounds. It is to be distinguished from *stammering* (*dysarthria literalis*) by the fact that in the latter spastic muscle contractions are absent. In stuttering the trouble with speech often disappears when the words are sung. Under observation stuttering is exaggerated, while stammering is usually somewhat lessened. There is a form of *syllable stumbling* which is met with especially in dementia paralytica.

In studying *aphasic disturbances* it must be kept in mind that speech consists of two great groups of functions, the *perceptive* (understanding of speech) and the *expressive* (act of speech). The anomalies on the perceptive side are included under the general term of *amnesic* or *sensory aphasia* (Wernicke). The memory for words can be disturbed in various ways; thus, the memory of names may be wholly or partially lost. The disturbance in which a patient hears spoken words but does not recognize their meaning is known as *word deafness*.

<sup>1</sup> For the literature of the subject and for excellent accounts the reader is referred to von Monakow (C.), *Gehirnpathologie*, second edition; Dejerine (J.), *Sémiologie du système nerveux* in Bouchard's *Traité de pathologie générale*, Paris, 1901, vol. v; and Moutier (F.), *L'Aphasie de Broca*, Paris, 1908.



When the patient whose speech muscles are not paralyzed has certain words and syllables in his consciousness but is unable to give expression to them in speech, he is said to suffer from *motor aphasia*. Such patients also are unable to speak words pronounced before them. They are never entirely dumb, being always able to give expression to a few words or parts of words; indeed, there is every gradation from syllable stumbling to almost complete dumbness.

Similar to aphasia are the disturbances known as *agraphia* and *apraxia*. In *agraphia*, although there is no actual paralysis of the muscles of the upper extremity, the patient is unable to write down words which he hears or sees or remembers. Usually *agraphia* is combined with aphasia, but in pure *agraphia* it exists by itself without aphasia. In *apraxia* (or *dyspraxia*) the patient is unable to carry out from memory certain complicated movements of his limb muscles. *Apraxia* seems to be due most often to lesions of the left hemisphere or of the corpus callosum.

The incapacity to understand written or printed matter is known as *alexia*.

When all the components of speech (perceptive and expressive) are interfered with, the condition is known as *total aphasia*. When the speech anomaly is chiefly on the expressive side, we designate it a *motor aphasia*. In the most common form of motor aphasia, that known as *Broca's type*, speech is almost abolished and the patient is unable to pronounce words spoken before him. He cannot read aloud, he has difficulty in spontaneous writing, in writing to dictation, and in copying. He may, however, understand words which he hears and sees. In cases of this type, in which the *agraphia* is much more pronounced than the speech disturbance, the special term of *cheirokinesthetic agraphia* has been applied.

In so-called *pure motor aphasia* (*subcortical motor aphasia* or *aphemia*; *pure word dumbness*) there is almost complete inability for spontaneous speech, for pronouncing words spoken before the patient, and for reading aloud, but there is no disturbance of writing and the understanding of speech and of writing is normal. In sensory aphasia, it is the perceptive side of speech which is interfered with. Here, again, we distinguish ordinary sensory aphasia (Wernicke's type) from pure word deafness.

In *ordinary sensory aphasia of Wernicke's type* (*cortical sensory aphasia*) the patient is unable to understand what he hears and reads. His storehouse of spoken words is somewhat diminished and his speech is *paraphasic* in that instead of using the words which he intends to employ he gives expression to others which have a similar sound. He is unable to pronounce words spoken before him, or if able to do so does not understand their meaning. The emphasis he puts upon spoken words is faulty. On spontaneous writing and on writing to dictation he manifests a verbal *agraphia* or mixes up the letters in words (*paragraphia*). He copies only with difficulty.

In so-called *pure word deafness* (*subcortical sensory aphasia*) internal speech may be normal, there is some deafness, and the patient is totally unable to understand words spoken to him, nor can he speak words pro-

nounced before him. He may, however, be able to express himself quite well in writing and to understand what he reads.

At the beginning of the sensory aphasias, especially in tumors or after brain injuries, patients sometimes pass through a transitory state in which motor speech is undisturbed, but there is difficulty in finding the names for objects and persons (concrete substantives). Such patients understand spoken and written words perfectly well, but on spontaneous writing they have the same difficulty as on speaking, although writing to dictation and copying may be quite normal. This particular condition is sometimes spoken of as *word amnesia* or *anomia*.

Two kinds of *pure word blindness* (*subcortical alexia* of Wernicke) have been described, one without *agraphia*, the other with *agraphia*. In *pure word blindness without agraphia* spontaneous speech and the understanding for spoken words are normal, but single letters and syllables cannot be read. The patients may write, but fail to understand the words that they have written themselves. They can write to dictation, but cannot copy. The condition is usually associated with *hemianopsia dextra*.

In *pure word blindness with agraphia* spontaneous speech and the understanding of spoken speech are normal, but there is complete *alexia* combined with *agraphia* and *paragraphia*, these being optic in their origin. The patient cannot write to dictation nor copy the writing of others.

**Anomalies of the Reflexes.**—A study of the disturbances of reflex action is of great importance in neurological and psychiatric diagnosis. Three main groups of reflexes should be examined: (1) The pupillary reflexes. (2) The deep reflexes (tendon and periosteal reflexes). (3) The superficial reflexes (cutaneous and mucosal reflexes).

**Pupillary Reflexes.**—These include (1) the light reflex, and (2) the accommodation and convergence reaction.

In testing the *light reflex* one must avoid calling forth movements due to accommodation or convergence. Each eye should be tested for itself, and one should also see whether or not the consensual reflex is present, remembering that on unilateral illumination there should be a pupillary contraction in both eyes.

In testing the *reaction on convergence* one asks the patient to look first at the ceiling and then quickly at the end of the nose. When the eyes converge for near vision there is also a contraction of the pupil due to accommodation. But even when the medial recti are paralyzed and convergence is no longer possible, there may be an accommodative pupillary contraction.

If the light reflex is abolished or diminished the condition is known as *reflex pupillary rigidity*. This may be due to partial or complete blindness (*amaurosis* or *amblyopia*) or to paralysis of the M. sphincter iridis; in other words, it may depend upon the centripetal fibres of the reflex arc (*nervus opticus*) or upon the centrifugal fibres (*nervus oculomotorius*).

When the pupils do not react to light but do react well on accommodation and convergence, we speak of the *Argyll-Robertson pupil*. Its presence is of very great significance in the diagnosis, especially of *tuberculosis* and of *dementia paralytica*. The condition is usually bilateral, but may in early stages be unilateral. When the pupils react neither to light

nor accommodation (*absolute pupillary rigidity*), there is complete ophthalmoplegia interna. This condition is not uncommon in cerebral syphilis, and may be met with also in brain tumor, tabes, or dementia paralytica.

The size of the pupils varies a good deal in health with moderate illumination. Abnormal contraction of the pupils (*myosis*) occurs in opium poisoning and in various conditions which irritate the oculomotor nerve or paralyze the sympathetic nerve. It should be borne in mind that myosis occurs physiologically in old people and in sleep. Abnormally large pupils (*mydriasis*) are seen in atropine and cocaine poisoning. The condition may also be due to oculomotor paralysis, to atrophy of the optic nerve, or to irritation of the cervical sympathetic.

Inequality in the size of the pupils (*anisocoria*) may be due to unequal illumination or to differences in the refractive media of the two eyes, but most often it indicates unilateral nervous disease of the optic, oculomotor, or sympathetic nerves.

**Deep Reflexes.**—Of these, by far the most important are the knee-jerk, the heel-jerk, and the periosteal reflex.

The *knee-jerk* (*patellar tendon reflex*, *knee-kick*) is the contraction of the M. quadriceps femoris which follows tapping upon the ligamentum patellæ. Sometimes the so-called *reinforcement* of Jendrassik is necessary; the patient clasps his hands and is told to pull at the moment one taps the patellar tendon. As a rule, it is more satisfactory to make the examination in the recumbent position.

*Achilles-jerk or Foot-jerk.*—The patient is placed on his knees in a chair, the feet hanging loosely over the end of the chair. One then taps upon the Achilles tendon with a percussion hammer. The reflex is not always present, even in healthy people, although generally so. If the Achilles jerk be exaggerated the percussion stroke may give rise to clonic contractions or an actual foot clonus instead of to a single contraction.

*Periosteal Radial Reflex.*—On tapping the lower end of the radius one can see a contraction of the M. brachioradialis at the bend of the elbow, leading to flexion and slight pronation of the forearm and hand.

The other deep reflexes which may be tested are: (1) *Tibial reflex* (striking anterior surface of tibia to get contraction of M. quadriceps femoris); (2) the *biceps reflex* (tapping the biceps tendon at the bend of the elbow); (3) the *triceps reflex* (holding arm at a right angle and tapping on the triceps tendon); (4) the *jaw-jerk* (laying finger above chin with the mouth closed and giving slight tap with percussion hammer to get contraction of the masseters).

The deep reflexes may be increased or diminished. An increase may be due to irritation of the sensory limb of the arc (neuritis, meningitis), to stimulation of the anterior horn cells (strychnine poisoning) or to diminution of inhibitory influences acting from above upon the reflex arc (neurasthenic states, lesions of the pyramidal tract). The deep reflexes may be diminished or abolished through injury to the sensory or motor nerves or injury to the portion of the arc within the central nervous system.

**Cutaneous Reflexes.**—The three principal ones to be considered are: (1) The plantar; (2) the cremaster; (3) the abdominal reflex.

The *plantar reflex* is elicited by applying a stimulus to the sole of the foot. Under normal conditions it leads to an involuntary contraction of certain muscles of the lower extremity, the so-called "tickle response" being a kind of movement away from the irritating object. Most important, however, is the behavior of the toes, and especially of the great toe. Under normal conditions plantar stimulation is followed by plantar flexion of the toes. On the contrary, when the pyramidal tract is injured, instead of plantar flexion there is dorsal flexion, especially of the great toe, and the movement of the toe occurs less rapidly than under normal conditions (*Babinski's phenomenon*). In children during the first few months of life, plantar stimulation causes dorsal flexion of the great toe, but after the first few months of life the normal reflex is one of plantar flexion. When the pyramidal tract is diseased the dorsal flexion of the great toe is often accompanied by spreading of the other toes, and especially by abduction of the little toe. This has been called the *fan sign*, (*signe de l'éventail*). Similar in its meaning is the response obtained by rubbing the medial surface of the tibia downward. Normally as the malleolus is approached, this causes plantar flexion of the toes, but in spastic states there results, as a rule, dorsal flexion of the foot and especially of the great toe (*Oppenheim's sign*).

Still another sign of similar import may be mentioned. If one taps the lateral part of the proximal half of the back of the foot, corresponding to the base or middle of the third and fourth metatarsal bones, the cuboid bone, and the second cuneiform bone, dorsal flexion of the toes occurs under normal conditions, while in spastic states we meet with a plantar flexion, sometimes with spreading of the toes (*Mendel's sign*).

**Cremaster Reflex.**—This is elicited by stroking the medial surface of the thigh in the adductor region or by pinching the skin in this location. The normal response consists in a contraction of the cremaster muscle, with elevation of the testicle. This reflex is not to be confused with the scrotal reflex, which consists of a contraction of the tunica dartos with wrinkling of the skin of the scrotum on stimulation of the skin in this neighborhood.

**Abdominal Reflexes.**—One strokes the skin of the abdomen with the end of the finger or with the blunt point of some instrument, and notices the contraction of the abdominal muscles which follows. It may be elicited either in the supra-umbilical or in the infra-umbilical region of each side (*epigastric and hypogastric reflexes*). A unilateral absence of the reflex is of most importance.

**Other Cutaneous and Mucosal Reflexes.**—Among the other reflexes may be mentioned the *palmar reflex*, *scapular reflex*, and *uvular reflex*. Especial mention perhaps should be made of the *lid reflex* (closure of the lids when an object is brought suddenly near one eye) and the *conjunctival* and *corneal reflexes*.

The reflexes set free by stimulation of the skin and mucous membranes have an entirely different significance from the deep reflexes. The cutaneous reflexes may be absent when the peripheral nerves are diseased; indeed, whenever the reflex arc is interrupted.

One of the most interesting facts connected with the cutaneous reflexes

is that they are especially disturbed in unilateral cerebral disease. Aside from the Babinski phenomenon, a hemiplegia usually leads to loss of the abdominal and cremaster reflexes on the paralyzed side, perhaps owing to an increase of the influences which inhibit the activity of the reflex arc, although this explanation is not universally accepted.

**Anomalies of the Vasomotor, Secretory, and Trophic Functions.**—

The vasomotor functions of the nervous system are very complex, and disturbances in different parts can lead to vasoconstriction on the one hand, or to vasodilatation on the other. For a full discussion of these vasomotor phenomena the reader is referred to the lecture by Porter,<sup>1</sup> the article by Eulenburg-Landois, and to the treatise of Cassirer.<sup>2</sup> Here belong the angioneuroses, acroparesthesias, angioneurotic œdemas, symmetrical gangrenes, erythromelalgias, acrocyanoses, etc.

Trophic disturbances may concern the muscles, the bones and joints, the skin, and its appendages. The trophic changes in the bones and joints have been best studied in tabes, in syringomyelia, and in acromegaly. In tabes spontaneous fracture of the bones frequently occurs, and still more frequently certain arthropathies, which occur suddenly, usually without pain, and lead quickly to disintegration of the joints. Trophic changes which take place in the skin include bed-sores (decubitus), falling of the hair, nails, and teeth in tabes, perforating ulcer of the foot in tabes and syringomyelia, and the panaritium of the finger tips in syringomyelia. Scleroderma and progressive facial hemiatrophy also belong here. Disturbances of the secretion of the sweat glands and of the sebaceous glands of the skin are sometimes met with in nervous disorders. The anomalies of the secretion of saliva, of the stomach juice, of the urinary secretion, etc., should also be mentioned.

<sup>1</sup> *Vasomotor Relations, The Harvey Lectures*, 1906-1907.

<sup>2</sup> *Die vasomotorisch-tropischen Neurosen*, 2nd ed., Berlin, 1912.

## CHAPTER II.

### DISEASES OF THE MOTOR TRACTS.

By WILLIAM G. SPILLER, M.D.

#### PROGRESSIVE SPINAL MUSCULAR ATROPHY.

**Synonyms.**—Muscular atrophy, type Duchenne-Aran; chronic anterior poliomyelitis.

**Definition.**—By the Duchenne-Aran type of muscular atrophy is meant a progressive muscular wasting related to degeneration only of the cells of the anterior horns of the spinal cord and their peripheral processes. While it usually begins in the small muscles of the hand, it may begin in the muscles of the shoulder. It is usually bilateral, but it may be unilateral for a time.

The name of Aran is sometimes placed before that of Duchenne and sometimes follows it. A paper was published by Duchenne in 1849, and it was not until a year later that Aran's work appeared. A further study by Duchenne was made in 1853. Both Duchenne and Aran looked upon the atrophy as muscular in origin. The anatomical investigations of Cruveilhier led to the disease being regarded as atrophy of the anterior roots. Luys demonstrated degenerative changes in the cells of the anterior horns. These and the investigations of Charcot, Joffroy, and others led to the conclusion that all progressive muscular atrophy was of spinal origin from disease of the cells of the anterior horns, a view that later was shared by Duchenne, although it was contrary to that he had previously held. The confusion at that period was very great, nor can it be said at the present day that complete order has been brought out of the chaos. Duchenne included all progressive muscular atrophy, whether occurring in families, in young or old persons, or as an acquired process, under one head. Gradually different types of muscular wasting were separated from the Duchenne-Aran form. Progressive muscular dystrophy in the pseudo-hypertrophic form had been observed by Duchenne in 1853. He at first erred in believing the pseudo-hypertrophic muscular paralysis was of cerebral origin, as his earlier cases manifested feeble intelligence, but later he recognized his mistake. He called the disease *paralysie musculaire pseudo-hypertrophique*, and this name has clung to it to this day. Erb established the unity of the various types of myopathy, but at first only the pseudo-hypertrophic and the Leyden-Möbius types were recognized. Charcot separated the amyotrophic lateral sclerosis from the general group of atrophy. Charcot and Joffroy distinguished the cervical hypertrophic pachymeningitis in 1871 to 1873, although it cannot be said that the symptoms of this disease are so sharply defined as are those of amyotrophic lateral sclerosis. Schultze and Kahler,

in 1888, weakened the Duchenne-Aran type of muscular atrophy by separating the great group of syringomyelia. It began to look as though nothing would be left in the Duchenne-Aran type, and indeed it is stated that Charcot himself felt that it stood on an insecure foundation.

In 1897, Marie came forth in defiance of the accepted teaching. He pointed out that Duchenne at first believed that progressive muscular atrophy was of peripheral origin; later, he believed it to be of spinal origin. The disease at first, as understood by Duchenne, included all the forms of muscular atrophy not resulting from fracture. Marie then described how amyotrophic lateral sclerosis, progressive muscular dystrophy, multiple neuritis, and syringomyelia were separated from the great group of atrophies. Fully one-third of Duchenne's cases, from his own description, Marie thinks, belonged to syringomyelia, although this disease at that time was not recognized. Thus, Marie says that the progressive muscular atrophy of Duchenne-Aran, which earlier authors regarded as the most solid foundation of neuropathology, has ceased to exist. Even the hand which is commonly regarded as of the Duchenne-Aran type is, according to Marie, not the deformity Duchenne described, and is what Duchenne believed to be characteristic of leprosy. The hand that he described as typical of his progressive muscular atrophy Marie believes belongs to syringomyelia. The atrophy is not so intense, and the fingers are not so much flexed; and yet in comparing the two hands, as pictured by Marie, the differences do not appear very great. Marie believes that Duchenne merely described a symptom complex, and that nothing now remains of his progressive muscular atrophy. This view is not generally accepted.

**Etiology.**—*Pregnancy* may have some influence on the development of spinal muscular atrophy, though it would seem questionable whether it be able to originate it when no tendency to the disease exists. In Oppenheim's second case of chronic poliomyelitis, a complicated case, weakness of the muscles of the right shoulder and of the right upper limb developed during the fourth pregnancy; the following year the left upper limb became affected during pregnancy. Nonne attributed the poliomyelitis in his second case to diabetes.

*Heredity* does not play a rôle, and yet Bruining has observed chronic anterior poliomyelitis in father and son, and in one of the cases obtained a necropsy; this family occurrence was probably merely a coincidence.

*Age* seems to be the most important factor, the symptoms usually appearing in middle life. The infantile form of progressive spinal muscular atrophy is considered separately. Myelopathy is probably an abiotrophy, and is the result of an imperfectly developed motor system unable to withstand the stress of advancing years.

It is exceedingly questionable whether infections or intoxications cause the symptoms, although the development of progressive muscular atrophy many years after an arrested acute poliomyelitis is regarded by Léri as evidence of this origin. C. S. Potts has paid particular attention to muscular atrophy occurring in this way, and collected the records of 36 cases. In 28 the condition was probably progressive muscular atrophy; in 2, amyotrophic lateral sclerosis; in 2, myelitis; and in 4, another

attack of acute poliomyelitis. The interval elapsing between the primary and secondary attacks ranged from seven years to about fifty-five years, the average being about twenty-three years. In 18 cases out of 33, in which the part secondarily affected could be determined, the late atrophy began in a limb which had previously been affected by the primary disease; in 7 it began in the other limb on the same side that had been primarily affected; in 5 in the corresponding limb of the side opposite to the part first affected; in 2 in a limb of the opposite side not corresponding to the limb first affected; and in one the involvement was general, this being a case in which acute poliomyelitis occurred a second time. The nerve cells left weakened by acute poliomyelitis may degenerate later, but this is not a proof that the chronic form is so caused. Some toxic substance acting on the cells weakened by acute poliomyelitis may cause progressive muscular atrophy. It seems probable that lead may be one of these agents. The acute and chronic forms probably have a very different pathology. The male sex is regarded by certain authors as much more frequently affected than the female.

Some of the cases supposed to represent progressive spinal muscular atrophy are caused by myelitis, especially the syphilitic form. The inflammation may be almost confined to the gray matter, and such being the case, sensory disturbance may be absent. The symptoms then would be those of progressive spinal muscular atrophy, although the lesion would be a diffuse process of inflammatory character.

**Pathology.**—The characteristic changes are atrophy and degeneration of the nerve cells of the anterior horns of the spinal cord, and also in some cases of the motor nuclei of the medulla oblongata. Sometimes many of the nerve cells entirely disappear. Numerous hemorrhages have been observed in the gray matter in some instances (Bielschowsky, Spiller), extending even throughout the cord. The motor roots are atrophied, and contain fewer fibres than normal. The white matter of the spinal cord remains unaltered in a typical case, or at least the sclerosis is not in the area occupied by the crossed pyramidal tracts. It is questionable whether those cases in which a very slight degeneration of these tracts occurs, detectable only by the Marchi stain, should be classed as progressive spinal muscular atrophy or amyotrophic lateral sclerosis. These symptoms and the findings in the anterior horns may indicate that the lesions of the anterior horns have been of long duration, while the degeneration of the lateral columns evidently is very recent, and no clinical evidence of the slight degeneration of the pyramidal tracts may have been detected during life. It seems best to regard such cases as belonging to progressive spinal muscular atrophy or chronic poliomyelitis, but as marking a transitional stage to amyotrophic lateral sclerosis.

Beevor, in disputing the correctness of Gowers' view that amyotrophic lateral sclerosis and progressive spinal muscular atrophy are the same disease, stated that he (Beevor) had a case in which the symptoms began with atrophy of the small hand muscles, and later the shoulder muscles became affected, the lower limbs were only slightly implicated. Rigidity or increase of the deep reflexes was not present. Atrophy of the cells of the anterior horns was found, but the lateral columns were intact.



Degenerative changes in the anterolateral columns may be recognized as belonging to anterior poliomyelitis and progressive spinal muscular atrophy, provided the pyramidal tracts are not affected. It must be accepted that certain of the column cells may degenerate as well as the cells of the anterior roots. When the pyramidal tracts become implicated the cases are on the border line of amyotrophic lateral sclerosis. Perivascular round-cell infiltration has been seen (Bielschowsky).

It is uncertain whether the cellular changes are primary or secondary. In some instances they may be primary, in others they may be the result of inflammation in the surrounding tissues. Where the process has advanced so far that most of the cells have disappeared, the neuroglia of the anterior horns appears denser and the few nerve cells that remain show much pigmentation. The cellular destruction is usually greater in the cervical swelling; and the anterior roots usually are much atrophied. The peripheral nerves and muscles show more or less alteration.

Syphilitic myelitis probably is the lesion underlying a certain number of the cases of progressive muscular atrophy. Progressive ophthalmoplegia may become associated with degeneration of the motor cells of the medulla oblongata and spinal cord, as in two cases observed by Dana.

It seems to Dana an unnecessary confusion of symptomatology to transfer the diagnosis of progressive muscular atrophy of the Duchenne-Aran, or other type, to that of amyotrophic lateral sclerosis so soon as a little spasticity begins, especially as there may be nothing peculiar in age, course, or duration. The atrophy may start in the hands and ascend to the shoulders before there is any evidence of lateral sclerosis. The disease may then become arrested, the spastic symptoms disappear, and the patient may again present the type of Duchenne-Aran atrophy without any spastic symptoms. All that we can do, in the writer's opinion, is to acknowledge that the lateral columns may be slightly affected without producing clinical manifestations, but because we cannot always recognize this slight degeneration clinically is not sufficient reason to decline to make distinctions in the pathological forms of spinal muscular atrophy. It is a question whether we are solving the problem by following Dana in limiting the use of the term amyotrophic lateral sclerosis to those cases which show only from the beginning and dominantly the spastic and contracturing type of progressive muscular atrophy. Why must degeneration of the lateral columns develop previously to or simultaneously with disease of the cells of the anterior horns, and not later?

**Symptoms.**—The symptoms of chronic anterior poliomyelitis and progressive spinal muscular atrophy vary in typical cases. In the former the paralysis is supposed to develop within a few days or a few weeks, the atrophy appears later, and entire muscles or groups of muscles are paralyzed, the course of the disease is more rapid, and the paralysis develops first in the muscles of the lower limbs or shoulders. In progressive spinal muscular atrophy, the paralysis is proportional to the atrophy, one muscle fibre after another is affected, and the course is longer than in poliomyelitis. These distinctions are not always regarded, and some of the reported cases of so-called poliomyelitis might with greater right be

regarded as examples of progressive spinal muscular atrophy, or even as transitional forms of amyotrophic lateral sclerosis.

Dejerine and Thomas, who describe the disorder under the name of chronic anterior poliomyelitis, say that *atrophy* is the primary and essential symptom, and *paralysis* is secondary and proportional to the atrophy. Thus, according to these authors, one of the most important diagnostic points between chronic poliomyelitis and progressive spinal muscular atrophy disappears. Beevor made no sharp distinction between the two processes, and remarked that wasting may begin in the lower limbs.

The first indication of disturbance may be awkwardness in the use of one or both hands. Usually one is affected before the other. Abduction and apposition of the thumb with the fingers become affected, the interossei and lumbricals waste, and the hand becomes much atrophied, including the thenar and hypothenar eminences. When the thenar eminence is wasted the hand is of the simian type. Later, when the interossei and lumbricals are attacked the flexion of the phalanges upon the metacarpal bones and the extension of the phalangeal articulations become impaired, so that the first phalanges are extended and the second and third flexed, the palmar tendons are prominent (*main en griffe*). The *atrophy* gradually extends to the flexors on the forearm, then to the extensors. According to Beevor, the flexors of the fingers and thumb are affected before the flexors of the wrist. The atrophy reaches the deltoid and muscles of the upper part of the limb and shoulder girdle. Beevor states that the triceps, latissimus dorsi, and the lower half of the pectoralis major usually escape. The trunk and neck muscles waste and the ribs become prominent; the lower limbs likewise become greatly atrophied. Dejerine and Thomas dispute the extension of the process to muscles innervated by bulbar nerves, as no necropsy has demonstrated such an extension, and they assert that the face is always intact, and Dejerine believes that bulbar palsy belongs to amyotrophic lateral sclerosis. Beevor, however, speaks of bulbar paralysis as a part of chronic anterior poliomyelitis, and Léry also mentions atrophy of the muscles of the face, although he adds that the condition is very different from that of bulbar palsy.

There are a number of cases recorded in which the atrophy began in the lower limbs, especially in the peroneal distribution, and in some instances (Moleen and Spiller) the weakness and atrophy developed rapidly. The upper limbs became implicated later. In other cases the muscles of the shoulder girdle are first affected (Vulpian, Dejerine) or the extensors of the fingers and wrists, as in a case reported by C. S. Potts. The muscles of the trunk may be first affected.

The *weakness* and *atrophy* probably begin more commonly in the hands, because the movements of these parts are highly differentiated; it is not probable that the weakness is first noticed in muscles employed frequently in specialized movements simply because the interference with these movements is more striking to the patient, and that in reality other muscles are affected at the same time. Edinger's exhaustion hypothesis, that those muscles most used are most likely to be the first to show alteration, is a reasonable explanation for the commencement of the

symptoms in the hands in most cases. The paralysis is flaccid and the joints relaxed, producing the condition known as flail joint, especially noticeable at the wrists. Grunow and Loevegren go so far as to make the atrophy of the lower limbs the first sign diagnostic for poliomyelitis in distinction to progressive spinal muscular atrophy.

Fibrillary tremors are very common in the atrophying muscles; sometimes they implicate small bundles of muscle fibres, and then they may more properly be described as fasciculatory. These quick, wave-like movements are not present in the atrophying muscles at all times, but in most cases may be detected at some period or other. They may be so pronounced as to correspond to the condition known as "muscular madness" (*folie musculaire*), but they do not cause movement of a segment of the limb, unless possibly of a finger. They are an early sign, and disappear as the atrophy of the muscles becomes intense. They may be present at one period in spinal atrophy, but later disappear, so that the absence of fibrillary tremors by no means excludes a spinal origin.

The tendon reflexes of the affected limbs become diminished or lost, and may be lost before atrophy appears, the cutaneous reflexes become less active where the muscles are wasted. Exaggerated reflexes should make the diagnosis of progressive spinal muscular atrophy or chronic anterior poliomyelitis doubtful, but, according to certain authors, do not exclude this diagnosis. Thus Oppenheim does not regard exaggeration of the tendon reflexes as sufficient to make a case of muscular atrophy one of amyotrophic lateral sclerosis.

The electrical reactions are important in the diagnosis between the myelopathic and the myopathic forms of muscular atrophy. In the latter the irritability is diminished, but the formula is not altered; in the former the nerves may respond to the faradic and galvanic currents, although not in the same degree; later the response to either current may be lost. The muscles at first require a stronger current, then their faradic irritability may be lost; when the galvanic irritability is increased, the formula may be reversed, so that the anodal closing contraction is equal to or greater than the cathodal closing contraction (reaction of degeneration); still later all electrical irritability of the affected muscles may be lost. Especially valuable as a sign of myelopathy is the modal change, so that the muscles contract very slowly to the galvanic current. The entire muscle does not always show electrical alteration, and only a few fibres may present this change, while adjoining fibres in the same muscle are normal; this is because fibres much atrophied may be in juxtaposition to normal fibres.

Sensory changes are not a part of myelopathy. Objective disturbance of sensation does not occur in an uncomplicated case, and while pain may occasionally be complained of, it is of a peculiar kind; it is the pain from overuse of wasting muscles, and not that from irritation of sensory fibres. It is more a dull aching like that in fatigued muscles, and not sharp, such as is felt in toothache. Some authors speak of pain occurring in the beginning of myelopathy, but skepticism as regards the correctness of the diagnosis is justifiable when pain is intense.

The sphincters of the bladder and rectum escape, but theoretically one

may understand how they could become affected if the nerve cells of the conus were degenerated. Atrophy of bone has been described by some authors. Cutaneous lesions are probably epiphenomena. Vasomotor symptoms may be present, and the atrophied limbs may be cold.

*Chronic external ophthalmoplegia* is probably a form of progressive muscular atrophy of the central type. It is a progressive paralysis of the external ocular muscles. Numerous cases are recorded in the literature. It is usually a disease of infancy, but may develop later in life.

**Diagnosis.**—Remembering the pathology we understand why the symptoms are muscular atrophy, weakness, loss of reflexes, and changes in electrical reactions. The process is slow (progressive muscular atrophy) or in periods of exacerbations (subacute or chronic poliomyelitis).

*Progressive muscular dystrophy* is to be distinguished by the age at the onset, the involvement first of the proximal parts of the limbs, the hereditary or family tendency, the absence of fibrillary tremors and of reaction of degeneration, the presence of pseudo-hypertrophy, etc. *Amyotrophic lateral sclerosis* is clinically progressive spinal muscular atrophy or chronic poliomyelitis with exaggeration of tendon reflexes, and pathologically with degeneration of the cells of the anterior horns and peripheral motor fibres and the pyramidal tracts; the latter causes exaggeration of the tendon reflexes. *Multiple neuritis* is usually associated with spontaneous pain, objective sensory disturbances, and tenderness of nerve trunks. The onset is usually more rapid, and the symptoms may be confined to the distribution of certain nerves. *Acute poliomyelitis* is to be recognized by signs of infection and rapid development of symptoms. The paralysis at first may be extensive, but gradually certain of the affected muscles recover, leaving others permanently paralyzed. *Syringomyelia* has the dissociation of sensation, impairment or loss of temperature, and pain sensations, with much better preservation of tactile sensation; also trophic disturbances, and it may be more nearly unilateral. The *neurotic muscular atrophy* of Charcot, Marie, and Tooth may closely resemble chronic poliomyelitis, but where sensory disturbances are present the distinction is clear. It is likely to be hereditary, and is more common in males. The atrophy is confined to the peripheral parts of the limbs, even after many years. *Leprosy* may have some resemblance in that it may begin in the hands and extend toward the trunk in the upper limbs, implicate the lower limbs in a similar manner, and be associated with reaction of degeneration. The lepra bacillus in the tissues, the dissociation of sensation like that of syringomyelia, the local swellings in the nerves, and the history of contagion make the diagnosis possible. *Cervical hypertrophic pachymeningitis* may cause an atrophy of the hands, but there is a period of pain in the upper limbs with objective sensory disturbances.

**Prognosis.**—Arrest is possible but improbable. The weakness and atrophy are likely to extend until a large portion of the body is affected, or nerve cells concerned with respiration or controlling the heart, are attacked, or death occurs from some intercurrent disease, but usually the process is very slow, and in one of Dejerine's cases the symptoms lasted about sixteen years.

**Treatment.**—This is of little value. Electricity, especially in the form of the constant current, moderate exercise, and passive movements are recommended and may be useful. Tonics and exercise in moderation help to keep up the general strength, but nothing will restore the muscles atrophied from degeneration of their nerve cells, or probably have much effect in arresting the course.

### PROGRESSIVE SPINAL MUSCULAR ATROPHY OF CHILDHOOD, OF FAMILIAL OR HEREDITARY CHARACTER (TYPE WERDNIG-HOFFMANN).

**History.**—The first cases were described by Werdnig, who reported 2; Hoffmann followed, with a report of 20 cases; Thomson and Bruce, with 1 case; Bruns gave, in 1901, a description founded on the cases in the literature to that date. Hoffmann's cases occurred in three families; he examined 6 clinically, 2 anatomically. Both of Werdnig's cases were with necropsy, and occurred in the same family, the patients had the same mother but different fathers. In Hoffmann's third family the disease was evidently transmitted through the mother. Bruns' 3 cases came from three different families. The family character was evident in the first case, as two of the other offspring of the parents had died from the same disease. Nothing concerning family tendency could be determined in the second case. The third patient was the only one affected of four children. Batten has reviewed the literature of this disease. The cases of Werdnig and Hoffmann began between the sixth and ninth months; only a few of the children could stand before the disease began, and none could walk. Death occurred in the most rapid case in the eleventh month; in the slowest in the fifth year. Bruns' first patient could walk when about a year and three quarters old, and died in the fifteenth year. His third patient also could walk when about one year and one-quarter old, and was still alive in the twelfth year.

Hoffmann attempted to classify the cases of hereditary progressive spinal and bulbar muscular atrophy, and makes the following types:

1. Occurrence in early childhood. Implication first of the pelvic girdle and atrophy extending from there toward the ends of the limbs (Werdnig, Hoffmann).

2. The bulbar paralytic facial type of childhood (Fazio, Londe).

3. A Duchenne-Aran type (Strümpell, Gowers).

4. An intermediate form (Bernhardt), which Bernhardt regards as of spinal and bulbar origin.

It seems to the writer that the atrophy of Werdnig-Hoffmann may be regarded clinically as progressive spinal muscular atrophy, differing from the usual type seen in adults in the commencement within the first years of life, in the implication first of the pelvic girdle and portions of the limbs near the trunk, and in the strong hereditary tendency, a tendency very feebly manifested in the progressive spinal muscular atrophy of adults; but pathologically the disease is amyotrophic lateral sclerosis of childhood, differing from the type in adults by the absence of exaggeration of tendon reflexes, by which we may infer that the peripheral motor

segments are always involved first. The case of Thomson and Bruce shows that they alone may be affected.

**Pathology.**—The findings described by Hoffmann are: Symmetrical intense degeneration of all the peripheral motor neurones below the hypoglossus, including the accessorius, *i. e.*, degeneration or disappearance of the multipolar cells of the anterior horns, so that few are left in a transverse section; intense degeneration of the anterior roots of the spinal cord, less intense changes in the peripheral nerves and the intramuscular nerve endings; degeneration of the crossed and direct pyramidal tracts, and of a portion of the lateral fundamental columns of the cord, more intense in the upper thoracic and cervical regions, but not traceable above the pyramidal decussation. The muscles show simply atrophy in all stages without complete disappearance of fibres, without multiplication of the sarcolemma nuclei, and with very little lipomatosis. The alteration of the spinal cord is more intense than of the muscles.

**Symptoms.**—The description given by Bruns is as follows: The disease begins in early childhood, progresses slowly, without fever or convulsions. The children never learn to walk, or gradually lose the ability to walk if they learned before the symptoms began. They cease to move the lower limbs in bed and to hold the trunk erect. Standing becomes difficult, and is possible only in the beginning of the disease, and then only with support. Paresis and atrophy of muscles occur nearly simultaneously in the muscles of the pelvic girdle and trunk, implicating symmetrically the iliopsoas and quadriceps femoris, and later the upper limbs and neck muscles. The proximal portions of the limbs, shoulder girdle, and pelvic girdle are affected, and the atrophy and weakness diminish toward the hands and feet, although these parts do not entirely escape in the late stages, but their movements are better than would be expected from the atrophy. The erect position of the head and trunk is difficult to maintain; the child cannot lie down slowly, and raises itself with difficulty. The vertebral column is distorted because of muscular weakness. The feet are in the position of talipes equinus. The atrophy is *en masse*, and may be partly concealed by fatty tissue. Diminished electrical response or reaction of degeneration is observed in the paralyzed muscles. Fibrillary tremors are present in some cases, absent in others. The bulbar muscles are occasionally affected. Hypertrophy or pseudo-hypertrophy does not develop. The paralysis is flaccid, the tendon reflexes are absent, and contractures occur. There are no sensory or mental disturbances, or impairment of the sphincters of bladder and rectum. The course is rather rapid, and death results from paralysis of the muscles of respiration. The disease is progressive, the progress of the atrophy is from the parts near the trunk to the peripheral portions of the limbs, and the atrophy is entirely symmetrical. Hoffmann believes that fibrillary tremors do not occur, mental development is not arrested, and the muscles of face, tongue, and throat are not paralyzed.

**Diagnosis.**—The greatest difficulty is in regard to muscular dystrophy, and so true is this that Senator says of his two cases that when the children were seen dressed and their mode of sitting, lying down, and standing up (climbing upon the lower limbs as in muscular dystrophy)

was observed, the diagnosis of pseudo-hypertrophy seemed correct, but when the clothes were removed no hypertrophy was visible. Most important in the diagnosis were the presence of fibrillary tremors and diminution in the electrical response, and possibly a commencing reaction of degeneration in one case. The cases were without necropsy, and it seems questionable to which type they belonged. The type of Werdnig-Hoffmann shows a more rapid extension of the atrophy than occurs in progressive muscular dystrophy.

Congenital myatonia (congenital hypotonicity of the muscles) differs in being a congenital disorder, in the absence of heredity and family tendency, and in the much greater involvement of the trunk and portions of the limbs near the trunk at a very early age. The differential diagnosis may be difficult.

### AMYOTROPHIC LATERAL SCLEROSIS.

In this chapter amyotrophic lateral sclerosis is considered as distinct from progressive spinal muscular atrophy and from chronic or subacute poliomyelitis.

**Etiology.**—Amyotrophic lateral sclerosis is probably an abiotrophy. The individual is born with a certain potentiality of his motor system, but at middle age the strain becomes too great and degeneration begins. There is very little to warrant the opinion that the disease is of toxic origin. It is possible that lead may be a cause, or at least it may cause a symptom complex very closely resembling that of this disease, as shown by S. A. K. Wilson. This author has studied four cases in which chronic lead poisoning began with double wrist drop, and in three of these the small muscles of the hand were not affected until late in the process, and in the other they escaped so long as the patient was under observation. In all four cases the atrophy was progressive, and was associated with some exaggeration of the tendon reflexes. Babinski's reflex may be found in this lead palsy associated with a certain degree of spasticity and with ankle clonus. The electrical reactions may be altered quantitatively but not qualitatively, and sensation may be intact, although in one of Wilson's cases lancinating pains were present, and this occurrence in association with the Argyll-Robertson pupil led to a diagnosis at first of tabes. No necropsies have been obtained, and, therefore, it is not known whether lead may produce exactly the changes of amyotrophic lateral sclerosis.

It is not unreasonable to assume that lead poisoning may appear as a systemic disease of the nervous system, and there is even a possibility that it may assume the type of tabes dorsalis, as shown by a case reported by Redlich. The commencement of the paralysis in the extensors of the hands and fingers is unusual in amyotrophic lateral sclerosis, but does occur. Muscular pains are not uncommon in lead poisoning, and cramps sometimes are felt in amyotrophic lateral sclerosis. An Argyll-Robertson pupil is rare in the latter disease, but Wilson seems to have observed it three times.

The attempt to give causes for which there is no evidence is hardly a

justifiable procedure. We have no reason to believe that traumatism, exposure to cold and wet, or sexual excesses play a rôle.

Syphilis may cause the symptoms of amyotrophic lateral sclerosis although it is not probable that all cases are caused by syphilis. Syphilitic disease of the cord may produce spasticity, weakness of the limbs, atrophy of the small muscles of the hands, exaggerated tendon reflexes and fibrillary tremors. Careful examination, however, will usually reveal other symptoms; thus there may be severe pain in different parts, or disease of the optic nerves; or the history may mention a hemiplegia, perhaps transitory. The writer has studied such cases, in which many of the symptoms suggested amyotrophic lateral sclerosis.

**Frequency and Sex.**—Raymond and Cestan reported, in 1905, 18 cases with necropsy in the service of the former at the Salpêtrière; 13 were in men and 5 in women. In Collins' cases, 55 were males and 49 females. Probst, in 1898, found that of 53 cases the number was evenly distributed between males and females. Heredity and occupation do not seem to exert any influence.

The writer has reported 8 cases of primary degeneration of the central motor tracts with necropsy; 2 of these belong to the type of primary lateral sclerosis, as the nerve cells of the anterior horns of the spinal cord escaped, and in neither of these cases was muscular atrophy present. Amyotrophic lateral sclerosis seems to be a rare disease, about as rare or possibly more so than syringomyelia.

**Pathology.**—There is a degeneration of the pyramidal tracts, of the nerve cells of the anterior horns of the spinal cord, of the anterior roots, intramedullary and extramedullary portions; of the peripheral nerves, of the muscles, and sometimes of the nerve cells and fibres in the motor cortex. The corpus callosum and the posterior longitudinal bundles may be affected. It is essentially a death of the motor system and muscles.

The fibres that stain by hematoxylin in the pyramidal tracts of the spinal cord in this disease were noticed by Charcot and Marie to be very fine. Anton also observed an unusual number of fine fibres in the pyramidal tracts, and a similar observation was made by the writer, although fibres of larger size were mingled with these small fibres. The latter in the sclerotic tracts suggest that atrophy as well as degeneration of nerve fibres plays a rôle, but the Marchi method indicates very clearly that degeneration of the myelin sheaths is very important. The central motor tracts develop from above downward, and the terminal portions being the last formed are the least resistant, and, therefore, degenerate first.

The degeneration of the *pyramidal tracts* is not so intense in amyotrophic lateral sclerosis as in the sclerosis resulting from a cerebral lesion. This is probably because one fibre after another is attacked in the process and all do not begin to degenerate at the same time. There is some doubt whether the process in most cases is ascending in each nerve fibre attacked, and the mere cessation of the degeneration at a certain level is no proof of an ascending process. It is reasonable to believe that there is a gradual death of the portions of the central motor neurones most remote from the cells of origin, and that the extent of the degeneration varies in different cases, but that the whole portion that is affected in



each fibre is diseased usually almost simultaneously, although the individual nerve fibres are attacked at different periods.

Kojewnikoff was the first (1883) to trace degeneration of the pyramidal fibres from the lower end of the cord to the cerebral hemisphere. In the two hemispheres the "corps granuleux" (fatty granular cells) were in symmetrical areas, and in almost equal quantity on the two sides.

Charcot and Marie (1885) traced the degeneration to the motor cortex in two cases. Kojewnikoff (Koschewnikoff) in a second case found degenerated fibres throughout the central motor tracts, and did not depend merely on the presence of fatty granular cells. In Lennmalm's case the pyramidal tracts were said to be degenerated from the cerebral cortex throughout their extent. Lumbroso traced the degeneration to the cortex. Mott traced the degeneration through the inner capsule and into the motor cortex. Rossi and Roussy, in 1907, found 18 cases on record in which the degeneration had been traced to the cortex.

As regards the degeneration of the pyramidal tracts the writer has been able to trace degeneration of the tracts as high as the motor cortex in 1 case, as high as the internal capsule in 2 cases, as high as the cerebral peduncle in 2 cases, as high as the pons in 3 cases. The degeneration does not usually extend above the pons, and gradually disappears. The higher regions of the pyramidal tracts show a more normal manner of staining. In one case degeneration of the pyramidal tract was traced on one side as high as the pons, and on the other as high as the lower part of the internal capsule. The degeneration therefore is not always symmetrical on the two sides, although it usually is. The writer has observed great spasticity of the limbs when the degeneration of the pyramidal tracts was barely detectable. The weakness may be due entirely to implication of the central motor tract alone, but the writer has observed paralysis on one side greater when the degeneration of the pyramidal tract on the same side was less intense than that of the other side. A study of the nerve cells showed that the motor cells of the spinal cord on the side of the less affected pyramidal tract were the more diseased.

Marie has stated that the degeneration of the lateral columns involves a greater area than that occupied by the pyramidal tracts, as shown by secondary degeneration from cerebral lesions or by myelination of the cord. It is unquestionable that the degeneration does extend beyond the pyramidal tracts, and this alteration of the supplementary zone has been described in progressive spinal muscular atrophy by J. B. Charcot.

Pilez found degeneration by the Marchi method in the anterolateral columns much greater than in the pyramidal tracts. This would indicate that the fibres in the former were more recently affected, and the condition in this respect resembles the degenerative changes occurring in the spinal cord in cases of grave anemia. There is, however, another explanation. The fibres of the pyramidal tracts extend a long distance, and when they are degenerated the change is seen through a great part of these tracts, and has the appearance of long-standing alteration; whereas in the anterolateral columns the fibres are much shorter, and many more fibres must degenerate to cause a long-standing alteration as intense as that of the pyramidal tracts.

The anterior horns may contain fewer fibres than normal; the cells of the anterior horns may be much affected, in some the nucleus is displaced to the periphery, and the chromophilic elements entirely destroyed, except where they surround the nucleus, and the remainder of the cell may have a pale green appearance by the thionin stain. The pigment is sometimes much increased. In some cells the nucleus may disappear, in many the chromophilic elements are changed to fine granules. Many cells have no dendritic processes, or only imperfect ones; some of the cells are tumefied and rounded, some have altered cell bodies and normal dendritic processes.

The *bulbar* changes are often pronounced. The nuclei of the hypoglossal nerves may be much degenerated or even disappear, so that the cells may be few in number, with chromatolysis and much pigmentation, and loss of dendritic processes; or they may entirely disappear. The intramedullary fibres of the hypoglossus may be intensely degenerated. The tongue muscles may be greatly wasted, in association with the alteration of the hypoglossal nerve, and show excess of interstitial fatty tissue. The nucleus ambiguus, as well as the posterior nucleus of the vagus nerve, may be much degenerated, but as the cells of the former do not form a very sharply defined group, it is often difficult to determine a partial destruction of this nucleus. The soft palate may be wasted.

The motor nucleus of the trigeminal nerve has been found affected (Rossi and Roussy, and others), but may escape when other bulbar nuclei are diseased. The facial, the glossopharyngeal, and spinal accessory nerves, and their nuclei, may share in the alteration. The nuclei of the ocular muscles, according to Marie, are never affected in this disease, but Hoche has found degeneration of the oculomotor, trochlear, and abducent nerves, and this indicates that ophthalmoplegia might occur.

Bulbar symptoms may exist without distinct alteration of the bulbar nuclei. The microscopic examination of a case reported by Dercum and the writer showed that the bulbar nuclei were for the most part intact, but the degeneration of the pyramidal tracts extended above these nuclei. It is possible that nerve fibres controlling these bulbar nuclei were affected, and this would seem to support Charcot's view of the process as primarily a degeneration of the pyramidal tracts. Oppenheim observed symptoms of disease of the ninth, tenth, and eleventh nerves in amyotrophic lateral sclerosis without changes in the nuclei of these nerves.

The bulbar nuclei, on the other hand, may be affected without implication of the pyramidal tracts. Duval and Raymond have reported a case in which the bulbar nuclei were affected and no trace of sclerosis of the motor tracts was seen. Raymond refers to a number of other cases in which the nuclei were affected but the motor tracts escaped. Miura asserts, however, that no case of progressive bulbar palsy without degeneration of the pyramidal tracts is to be found in the literature.

It occurred to the writer in 1899 that inasmuch as amyotrophic lateral sclerosis is a disease of the motor system, a case in which the cortex was degenerated could be employed in delineating the extent of the motor area in man. The process seems almost like an experiment. This use of amyotrophic lateral sclerosis was original with the writer, and later

was adopted by others. Intense degeneration of the precentral convolution was found and considerably less degeneration of the postcentral convolution, and therefore it was concluded that the latter is also a part of the motor cortical area. The postcentral convolution may escape. It is proper to state, however, that the tendency is to exclude the postcentral convolution from the motor region.

Degeneration of the corpus callosum has been found by Probst, Spiller, Rossi and Roussy, and Naka in the middle portion of this structure. In the writer's case it extended in less intensity to the knee.

The large cells of the motor cortex have been found diseased. Mott found that many of them had disappeared. Marinesco has found the giant cells of the motor cortex greatly diseased. Most of these cells had disappeared, and the few remaining were atrophied, and had undergone chromatolysis and displacement of the nucleus.

Some degeneration of the columns of Goll in the cervical and upper thoracic regions has been observed in a case of primary lateral sclerosis reported by Dejerine and Sottas. Sensation had been normal. Moeli observed degeneration of these columns in these regions in a case of amyotrophic lateral sclerosis.

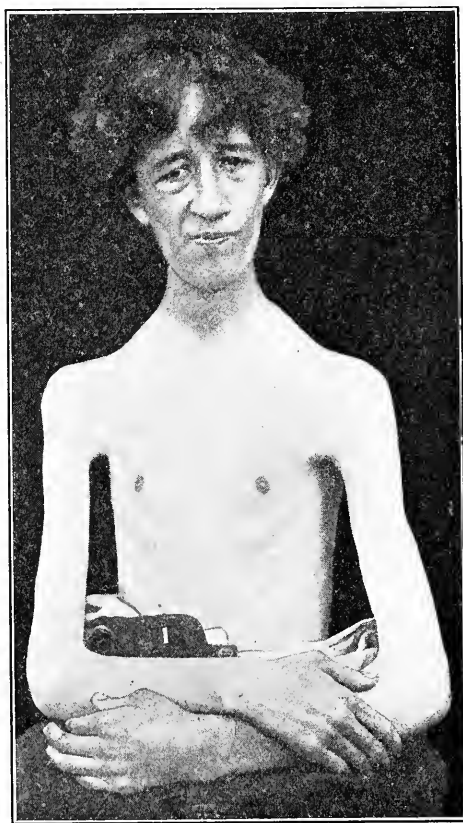
The columns of Goll are very liable to undergo a slight sclerosis in the lower cervical and upper thoracic regions in cases where no symptoms of this sclerosis have been present, and in some way it seems to be related to disease of the central motor tracts. Oppenheim, and Charcot and Marie have noticed sclerosis of these columns in amyotrophic lateral sclerosis. These cases must be distinguished from those in which the symptoms of posterior sclerosis were added to those of sclerosis of the lateral columns, a posterolateral sclerosis. Nonne observed a distinct though not excessive rarefaction of fibres in the median portion of the posterior columns in chronic anterior poliomyelitis; it was most evident in the cervical region. The degeneration of the posterior columns in these cases of primary degeneration of the motor system is not tabetic in character, it is not systemic. The posterior roots are not degenerated, and the region affected, lower cervical and upper thoracic, is not that usually diseased in tabes.

The muscles may be intensely atrophied, but even in many of these atrophied fibres the transverse striation may be distinct; in others the longitudinal striation may be more visible. The muscle fibres may or may not show fatty degeneration. In some of the fibres striation may not be clearly seen. The vessels may be much altered. Much fatty tissue may be found in some parts of the field. The nerve fibres between the muscular fibres may appear normal. The ulnar nerve has been found degenerated in one case (Spiller).

**Symptoms.**—This is a disease of middle life. Marie regards those cases (Erb, Seeligmüller) in which it is said to have begun in childhood as very doubtful, and believes that some of them belong to the family spastic paralysis. Marie gives the commencement between the ages of thirty-five and fifty years; Dejerine, the second half of adult life; Oppenheim, middle life; Eichorst, between thirty-five and fifty years; Gowers, between twenty-five and forty-five years; Strümpell, between twenty-

five and forty years. In Probst's statistics of 47 cases the disease began most frequently between thirty and fifty years, in 13 cases between fifty and sixty years, in 5 cases between sixty and seventy years, and in one case after seventy. Rossi and Roussi conclude that the beginning is usually between thirty and fifty, although frequently between fifty and sixty years; it is rare between sixty and seventy, and exceptional after seventy years. They have not been able to find any case recorded with

FIG. 1



Amyotrophic lateral sclerosis. With bulbar symptoms and greater implication of the proximal part of the upper limbs, in a boy, aged nineteen years. (Spiller and Gittings.)

a commencement after seventy years, except one case of Probst, but they report 2 cases from the service of Pierre Marie in which the commencement was at the seventy-first and seventy-third year.

Marie fixes the *duration* at eighteen months to two years, but death may occur in three to six months. Dejerine has observed a duration of nine years, and Florand of ten years. Marie questions whether these were truly cases of amyotrophic lateral sclerosis. Collins selected the records of 94 typical cases, and added to these 10 of his own but not all were with necropsy. He found that in 100 selected cases, 30 patients were between thirty and forty, 29 between forty-five and fifty, 28 between fifty and sixty, and 2 under thirty. In 26 cases the average duration was a little more than two years. In 2 cases the disease seems to have terminated in six months. Records of 4 cases were found in which the disease lasted only nine months. A few cases lasted only about five years; the longest on record was ten years.

Amyotrophic lateral sclerosis may begin occasionally at an early age. The writer saw with Dr. Gittings, a boy, aged nineteen years, who had marked bulbar symptoms with signs of degeneration of the nerve cells of the anterior horns in the cervical and thoracic regions, and possibly slight degeneration of the pyramidal tracts. The course was rapidly fatal, the symptoms being present only about one year.

The symptoms were first manifested in two of the writer's cases in one

lower extremity, although in one of these the upper limb of the same side may have been implicated at nearly the same time. The paralysis in these two cases became a hemiplegia and later a triplegia. In another case the paralysis seemed to begin as a hemiplegia. The statements of the patient or of the relatives must be taken in determining the part first affected, and there is, therefore, an element of doubt. The weakness and atrophy usually begin in the muscles of the hands, but sufficient cases are on record to show that they may commence in the feet. In a case reported by Dercum and the writer the weakness of the lower limbs preceded the bulbar symptoms about three years. In 81 cases collected by Collins, the upper extremities were first involved in 39 cases, the lower in 14 cases, the limbs of one side in the hemiplegic type, or all four limbs in 11 cases.

The writer collected reports of 11 cases (Dejerine, Pick, Vierordt, Leyden, Lennmalm, Mott, Senator, Probst (4)) in which the symptoms were confined to one side of the body, although both pyramidal tracts were affected in some of these cases. A case reported by C. S. Potts should be included in the list. Where the disease begins as a unilateral affection it does not remain as such, but sooner or later, and usually soon, the other side of the body becomes implicated. The shoulder muscles in rare instances may be affected first, and Dejerine and Thomas report two such cases, or the extensor muscles of the hand may be first affected.

Raymond and Cestan<sup>1</sup> from a study of their cases make four types of amyotrophic lateral sclerosis: (1) Ordinary spinal type; (2) labio-glossolaryngeal paralysis; (3) amyotrophic type; (4) spastic type.

1. **Ordinary Type.**—This corresponds to the classical descriptions of Charcot with spastic paraplegia or tetraplegia, ankle clonus, later amyotrophy, fibrillary tremors, and disturbances in the electrical reactions. Babinski's reflex was not observed, which seems to be remarkable. Only three of their cases corresponded to this type, and in two of them the amyotrophy began in the lower limbs and later implicated the upper limbs and the muscles innervated from the medulla oblongata. In one case the amyotrophy began in the hands. These cases lasted twenty-four, twenty-six, and twenty-six months respectively.

2. **Labio-glossolaryngeal Type.**—This was observed five times, four times in women, and Raymond and Cestan are inclined to think it is more common in this sex. The affection began in the muscles of the lips, tongue, and larynx, with exaggeration of the masseter reflex. Soon the limbs became implicated with exaggeration of the reflexes and progressive amyotrophy of the upper limbs. One patient had in the beginning attacks of choking, probably from spasm of the glottis; another had spastic laughing and weeping, without impairment of intellect. The cord showed degeneration in the anterolateral columns of the cervical and thoracic regions by the Marchi method. The duration of these five cases was from fourteen to twenty-four months.

3. **Amyotrophic Type.**—The disease appears as progressive muscular atrophy. The spasticity is slight, and ankle clonus and Babinski's sign are not obtained, but the bone and tendon reflexes are exaggerated, some-

<sup>1</sup> Raymond and Cestan, *Revue Neurologique*, 1905, p. 504.

times only slightly. The slight spasticity makes the diagnosis somewhat doubtful. The duration ranged from eight months to five years. The diagnosis must be made from the Duchenne-Aran type of muscular atrophy and subacute anterior poliomyelitis. The necropsies showed degeneration of the anterolateral columns. Nine of their cases were of this type. The disease of the peripheral motor segments is prominent in this type, but the implication of the central motor segments is indicated

FIG. 2



Amyotrophic lateral sclerosis. Intense atrophy of the hands.

by some exaggeration of the tendon reflexes, although spasticity is slight or even absent. It resembles subacute anterior poliomyelitis very closely, and, indeed, several of the cases of this latter disease with necropsy that have been reported belong to the non-spastic type of amyotrophic lateral sclerosis, as pictured by Raymond and Cestan.

4. **Spastic Type.**—Only one of Raymond and Cestan's cases belonged to this type. Spastic paraplegia was intense during one year, and was

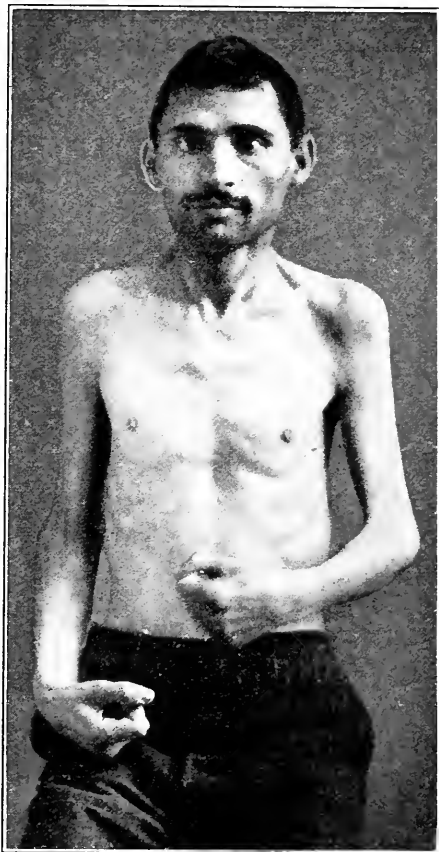
associated with ankle clonus and Babinski's sign, presenting the clinical picture of what is described by French writers as spastic dorsal tabes. Disseminated sclerosis, Erb's form of spinal syphilitic myelitis, and combined sclerosis in an early stage may present the same clinical picture. After a year progressive amyotrophy of the hands was observed, then the legs became affected, and still later the bulbar muscles.

Mills excludes Gowers' atonic variety in which both upper and lower limbs are the seat of wasting without spasm, except in cases in which the lower motor segments degenerate first, especially if the degeneration be complete, as any degeneration of the upper motor segments may then fail to produce the usual symptoms. It may be said, however, that all such cases clinically belong to progressive spinal muscular atrophy or chronic poliomyelitis, and their true relation cannot be recognized without necropsy. Not only may the peripheral motor segments be alone affected, but degeneration may be confined to the central motor segments.

The most important symptoms are weakness, spasticity, fibrillary tremors, exaggerated tendon reflexes, contractures, muscular atrophy, and bulbar disturbances. The *weakness* is an early manifestation, and is caused by the degeneration occurring in the central and peripheral motor tracts as well as by the atrophy of the muscles. It is usually first noticed in the hands; the patient loses the finer movements of the fingers and can no longer approximate the thumb to the little finger

with the same force, because of weakness in the muscles of the distribution of the median nerve. Sewing and writing become difficult. If he has learned any form of skilled movements, like playing on a musical instrument or typewriting, he has difficulty in performing these. Soon the wasting in the thenar and hypothenar eminences and in the interosseous spaces becomes pronounced, the weakness of the hand muscles progresses, and

FIG. 3



Amyotrophic lateral sclerosis beginning in the shoulder girdle and proximal parts of upper limbs. The utmost effort was made to elevate and flex the upper limbs, with little result. Flexion of the fingers was feeble, but far greater than movement in the rest of the upper limbs.

causes the simian hand, so called because of the loss of the thenar eminence. The extension to the forearm is gradual, and yet likely to occur early, while the arm and shoulder muscles usually are not implicated until considerably later. The weakness and atrophy increase *pari passu*, but the former is not dependent alone on the latter, as degeneration of the pyramidal tracts must cause weakness independent of any atrophy of the muscles.

The lower limbs are more or less *spastic* from the beginning, and weakness of them gradually develops. They do not, as a rule, present much atrophy until the disease has attained a considerable development; this is because the pyramidal tracts are early affected, while the cells of the anterior horns in the lumbar region may escape until late. Cases in which the symptoms begin in the lower extremities, and the upper extremities are not implicated until later, occur much more frequently than those in which the shoulder muscles are first affected. The muscles of the neck and trunk, as well as those of the face, may become greatly wasted, and the patient may be unable to hold the head erect.

The tendon *reflexes*, the biceps and triceps, von Bechterew's scapular reflex, the patellar and Achilles tendon reflexes are early exaggerated, and those of the lower limbs usually as much as those of the upper limbs. Clonus is common, both ankle and patellar clonus. It may even be seen in the upper limbs, as wrist, biceps or triceps clonus; but it is much less likely to occur in the upper than in the lower limbs. In 10 cases reported by Raymond and Cestan, in which the Babinski reflex was searched for, it was found only once, and that in an extremely spastic case. It is hard to understand why this sign should have been absent in the other cases, as extreme spasticity is not necessary for its production when the central motor tract is affected. The masseter reflex often remains normal when the tendon flexes of the limbs are exaggerated, but if the disease be intense, or extend far upward in the pyramidal tracts, the chin-jerk may be intensified as well as the other reflexes.

Beevor was the first to observe, in 1881, spontaneous clonus of the lower jaw. The sign, according to Beevor, shows that the sclerosis affects the pyramidal tract as high as the level of the fibres going to the motor nucleus of the fifth cranial nerve. He never observed clonus of the lower jaw in any other disease, except once in a case of hemiplegia. There may be some rigidity of the lower jaw with jaw clonus preceding the wasting of the bulbar muscles in the bulbar form of amyotrophic lateral sclerosis, and this may be absent in the true bulbar palsy.

In the case reported by the writer in which degeneration was traced to the cortex, there was some loss of power in the orbicularis palpebrarum of the left side, preventing the patient from closing the left eye tightly. The case is like one reported by Marinesco in the implication of the upper part of the face; and this is remarkable, as in bulbar palsy the upper part of the face usually escapes. The patient had a low-grade neuroretinitis and pronounced perivascularitis. The presence of this ocular condition does not exclude amyotrophic lateral sclerosis.

Fibrillary *tremors* occur usually in the atrophying muscles and may be intense, but may not be always present when the patient is under observa-



tion, and may disappear when the atrophy has attained an intense degree. They usually persist, however, a long time, and may precede the atrophy. They are indicative of degenerative changes going on in the cells of the anterior horns of the spinal cord, and have been described by the not very appropriate name of "fibrillary chorea."

The upper and lower limbs are usually spastic, but if the atrophy attains a considerable development the spasticity gradually diminishes and may disappear. It is not unusual to observe the spasticity greater in the lower than in the upper limbs, as atrophy is less likely to develop in the former. *Contractures* of the intensity seen in hemiplegia are rare, but there may be more or less fixity of the limbs from the increased tonicity of the muscles, and if this continue sufficiently long, contraction will occur and cause permanent fixity even after the spasticity has disappeared. Contractures are not very pronounced, but when present are usually of the flexor muscles. They may be so intense that the feet may be in the position of talipes equino-varus. In the upper limbs the arm may be held closely to the chest, the forearm flexed on the arm almost at a right angle, the hand flexed at a right angle to the forearm, the thumb turned inward and the fingers flexed. Involuntary jerking of the fingers is common, and may be related to fibrillary contractions. The wasting of the muscles and the gradual diminution in their tonicity diminish the contractures as the case progresses.

Marie has observed *intention tremor* of less intensity than in disseminated sclerosis. If such occur it probably is caused by muscular weakness and inability to coördinate properly the movements.

The *reaction of degeneration* is not usually very pronounced, as normal muscle fibres are mingled intimately with degenerated fibres, and the former mask the reaction presented by the latter. A change in the formula as well as a modal change may occur, but much more common is diminished response.

Retention of urine and loss of control over the bowels have been observed, and are probably the result of lesion of the pyramidal tracts.

A certain degree of *mental failure* may be observed occasionally, and the patients may take little notice of what is going on about them. Some writers have recognized mental deterioration as a part of the symptom complex. Marie says that mentality is always more or less affected, intelligence is enfeebled, emotionality is increased, and tendency to laugh or weep is pronounced. Sometimes a demented condition exists. The patient is moody, and intellectually and morally approaches the attitude of a child; at times the condition is like that of neurasthenia. These conditions are in association with bulbar symptoms. Suicidal impulses are not necessarily a sign of mental failure. One cannot wonder that the hopelessness of the disease should so impress the patient as to make death preferable to a miserable and prolonged existence.

*Pain* is sometimes observed. In one of the writer's cases the man complained of pain in both lower limbs below the knees five or six months previously. These were probably the pains of fatigue, as the patient was weak in the lower limbs and became very tired by night. Muscles undergoing atrophy and partially paralyzed may readily cause pain if

exercised beyond the limited power remaining in them. Naka observed pain in the beginning of the disease in his case, and no cause for it was found. He could not attribute it to the spasms, as Oppenheim has done, as his patient did not have spasms. Sensory disturbances of the objective form (anesthesia, hypoaesthesia, analgesia, hypoaesthesia) do not belong to the ordinary clinical picture, but pain on pressure has been observed. It seems reasonable to attribute these painful phenomena to an associated neuritis. Much caution is needed in making a diagnosis of amyotrophic lateral sclerosis when sensory symptoms are pronounced, as in most such cases some complication is probable or the diagnosis is incorrect.

The disease may begin as bulbar palsy, and when this type occurs death is usually too rapid to permit of much implication of the limbs. Or the bulbar implication may develop within a few months or a year or two after the limbs are affected, as in one of the writer's cases in which dysphagia and dribbling of saliva developed almost at the same time as the loss of power in the upper and lower limbs. The symptoms in this case lasted about one year when death occurred.

The muscles about the mouth become affected, the patient can no longer properly pucker up his lips as in whistling or in blowing out a candle, the lips sounds—l, b, t, r, s, k, g, ch, i, d, n—become imperfect, and fibrillary tremors become very distinct in the face and tongue. The upper part of the face, as in progressive bulbar palsy, usually is not affected. The tongue becomes deeply furrowed, and the atrophy and weakness may be so great that the patient may be unable to protrude it beyond the lips. The soft palate may be paralyzed and atrophied, so that fluids regurgitate through the nose when the attempt is made to swallow; the voice is nasal and very indistinct, and the soft palate no longer rises, or rises imperfectly when the attempt is made to say "a." The pharyngeal muscles also may be affected and swallowing becomes very difficult, and the pharyngeal reflex may disappear. The face may be much atrophied, death not occurring rapidly enough to prevent the atrophy, even when the bulbar nuclei are involved. Even when the lower part of the face is intensely implicated the eyelids usually can be held firmly closed. The vocal cords may be paralyzed.

Bulbar symptoms do not necessarily indicate a speedy fatal termination. In Raymond and Cestan's cases, in which the bulbar disturbances were the first symptoms, the duration of the disease was not shorter than in those in which the first symptoms were spinal, and bulbar disturbances developed later.

The muscles of the face, outside of those of the lips, usually escape. The muscles of mastication are not usually among the first affected in bulbar palsy, but they may be to such an extent that the patient is unable to chew or to move the jaw from side to side. Food may enter the larynx and cause attacks of coughing or aspiration pneumonia. Bulbar symptoms occurred first in 21 of the cases collected by Collins. In upward of 50 per cent. of all the cases, bulbar symptoms appeared within the first year. Marie is inclined to believe that labio-glossolaryngeal paralysis is always a part of amyotrophic lateral sclerosis.

**Diagnosis.**—What has been said in the differential diagnosis of progressive spinal muscular atrophy applies equally well to amyotrophic lateral sclerosis, when it is remembered that in the latter disease we have the symptoms of progressive spinal muscular atrophy with the symptoms of spasticity, exaggerated reflexes, and Babinski's sign.

**Prognosis.**—This is exceedingly grave. The disease is fatal, and may terminate within one year, and usually does so within three or four years. We have no means to arrest its course, and the end usually comes through the implication of vital centres in the bulbar palsy.

**Treatment.**—The section on treatment in the primary degenerations of the motor system need not be very extensive, as the results are not brilliant. The administration of strychnine may be of service. Over-exertion is to be avoided, as in this way too rapid exhaustion of the nervous system may be prevented. Massage, passive movements, and electricity are of doubtful value when the atrophy and weakness are progressive, although when the course is very slow or has been arrested they may be of great service.

#### **LATERAL SCLEROSIS (SPASTIC TABES, SPASTIC SPINAL PARALYSIS, SPASTIC DORSAL TABES).**

In a paper written in 1903, Erb remarks that he did not suppose when he first described spastic spinal paralysis in 1875 he would be compelled twenty-eight years later to fight for its recognition. His views expressed in 1875 received the support of Charcot and others, but met with the skepticism of prominent neurologists, especially of the Berlin school. The condition was regarded merely as a symptom complex caused by various disorders of the spinal cord and brain; as a condition without a definite pathology. Necropsies did not confirm the views of Erb and Charcot that primary degeneration of the pyramidal tracts was the underlying lesion, and while degeneration of these tracts was found, it was usually secondary to hydrocephalus, multiple sclerosis, tumor, syringomyelia, or symmetrical cerebral lesions; or was part of postero-lateral sclerosis or of amyotrophic lateral sclerosis.

Leyden and Goldscheider acknowledge the possibility of primary degeneration confined to the pyramidal tracts, but the clinical diagnosis of this condition they believe is impossible, and therein is the important factor. It is impossible, they think, to speak of an etiology, pathogenesis, and therapy of spastic spinal paralysis. Strümpell does not accept this view, but regards as the important factors the clinical and anatomical findings; when these are established the diagnosis will be easy. As justifying the recognition of spastic spinal paralysis, Erb accepts 11 cases; they are those of Morgan and Dreschfeld, Minkowski, Strümpell (2 cases), Dejerine and Sottas, Donaggio, Friedmann, Bischoff (2 cases), Demock, and Kühn.

**Pathology.**—This, if it is to be accepted as an entity, consists of primary degeneration of the pyramidal tracts and nothing else, and this degeneration may stop at any level or extend to the cortex. If we accept all

the 11 cases that Erb includes, we must be a little more liberal in our interpretation. Thus, in Morgan and Dreschfeld's case, atrophy of some cells of the anterior horns in the thoracic and lumbar regions was found, but Erb accepts it because this degeneration seemed unimportant and amyotrophy was not present during life. Grainger Stewart stated that he examined the sections of this case and found the changes in the gray matter distinct. At the time this case was reported Nissl's method was not in vogue, and could it have been employed it is not improbable that more evidence might have been obtained justifying the classification of the case as one of amyotrophic lateral sclerosis.

Implication of the direct cerebellar tracts, as in Minkowski's case, may be accepted as possibly causing no clinical signs, and therefore need not exclude a case from the designation of spastic spinal paralysis. Whether the same may be said of a slight degeneration of Goll's columns in the upper part of the cervical region, as in one of Strümpell's cases and in others, is open to question; but it occurs in certain cases of amyotrophic lateral sclerosis, in which no clinical evidence of its presence is observed. Strümpell, however, made the diagnosis in his case as combined system disease. Where syphilitic meningomyelitis exists the degeneration of the pyramidal tracts cannot be regarded as primary, and if a nuclear stain be not employed the cellular infiltration of the pia and cord may be overlooked.

**Symptoms.**—These, as originally described by Erb, are motor weakness, spasticity, and exaggerated reflexes; to these should be added the Babinski reflex. No other symptoms should be present, there should not be sensory disturbance, atrophy, sphincteric disorder, tumor, ataxia, or implication of the cranial nerves. The symptoms may persist many years, and Erb quoted cases in which they have lasted nineteen to twenty-seven years. They may develop very slowly, during twenty-three years in the case of Dejerine and Sottas.

The *weakness* usually affects one lower limb some time before the other lower limb becomes implicated, but it is often much less intense than the spasticity. It is at first a sense of fatigue after slight exertion, and spasticity is associated with the paresis. The patient stubs his toe in walking because of the spastic condition of the muscles of the lower limbs, and the manner in which the soles of the shoes are worn is often very diagnostic, the sole at the point of the shoe may be completely worn away. The Achilles tendon and patellar tendon reflexes are much exaggerated, so that ankle clonus and patellar clonus may be persistent, or the whole lower limb may be thrown into clonic movement, a condition inappropriately called spinal epilepsy by French writers. The lower limbs are rigid, the thighs adducted, so that walking is interfered with, and the scissors gait, so called because of the tendency for one limb to be drawn in front of the other, is produced. The spasticity may be so pronounced that marked spasm is caused by passive movement. If the leg be extended upon the thigh, passively, a sudden spasm may jerk the leg to full extension, the "clasp-knife" reaction.

The upper limbs may escape entirely, but if they be affected the symptoms may be less marked, though of the same type as those in

the lower limbs. In later stages of the disease the patient becomes bedridden and unable to stand. Death results from some intercurrent disease.

Slight flexion at the knees Strümpell believes will be found in all cases of spastic spinal paralysis beginning in early youth. This is usually attributed to spastic contracture in the affected muscles, but Strümpell believes it is caused by differences in growth of certain muscle groups. The flexors are more affected than the extensors, therefore their growth is arrested when the lesion develops early in life.

Strümpell distinguishes the following forms of spastic spinal paralysis:

1. A form certainly endogenous, characterized by family and hereditary appearance, males especially affected, commencing between the twentieth and thirtieth years, progressing very slowly, lasting a long time (twenty to thirty years or longer). The symptoms are gradually increasing rigidity and hypertonia of the muscles of the lower limbs, exaggeration of tendon reflexes, etc. Gait is spastic and for a long time without marked paresis, but paresis occurs later, especially in the flexors and muscles of the lower part of the lower limbs. The upper limbs remain intact until the end, likewise the bulbar and cerebral functions.

2. Infantile spastic spinal paralysis. This commences in childhood, at times as early as the third to sixth year, although it may begin later. This type is related to the cases of congenital abnormal development. A careful history must be obtained in order to decide whether the condition be congenital agenesis, or progressive degeneration beginning later. Inquiry must be made regarding injuries at birth, healed encephalitis, etc. The symptoms are those of spastic paralysis of adults, but modified by certain conditions of the developing organism. The upper limbs are often unimplicated, or at least only slightly so, but they may be distinctly affected. Arrest in cerebral development is often present, shown by mental deficiency. Such cases are not very rare.

3. A third type is also endogenous in origin. It begins in late life, progresses comparatively rapidly, implicates the entire pyramidal tracts, so that the upper limbs and bulbar muscles are affected. The symptoms are general spastic rigidity, peculiar spasticity in bulbo-cerebral territories (glottis spasm, forced phonation, forced laughter, forced crying, involuntary movements of the facial muscles, etc.). The tendency to implication of the peripheral motor neurones is strong, shown by fibrillary twitchings, muscular atrophy, etc. These cases are closely related to amyotrophic lateral sclerosis. They usually are isolated, not occurring in families.

4. An exogenous type. The syphilitic spinal paralysis, with the exception of lathyrismus, is the only important form. The lesions are seldom confined to the pyramidal tracts, and the columns of Goll are usually partially affected. The clinical picture of spastic spinal paralysis is seldom uncomplicated.

5. A fifth form may occur in women after confinement and the puerperium. Strümpell has observed a number of cases in which spastic spinal paralysis developed following these conditions. No necropsies of cases of this form have been obtained.

**Diagnosis.**—This is often difficult so far as the exclusion of other processes is concerned, and among these may be mentioned spinal tumor without pain, multiple sclerosis, hysteria, latent caries of the spine, myelitis, secondary degeneration from cerebral lesions, and combined sclerosis in which the symptoms of posterior column disease are indistinct. Indeed, it is questionable whether the diagnosis of primary lateral sclerosis can be made with absolute certainty during the life of the patient.

*Tumor of the spinal cord* is occasionally difficult to distinguish. In it the symptoms are usually unilateral at first, bilateral later, and associated with severe shooting pains, but cases of spinal tumor occur in which the symptoms are bilateral from the beginning and pain is absent. In a number of cases the diagnosis between spinal tumor and primary lateral sclerosis seems impossible.

*Multiple sclerosis* usually presents characteristic symptoms, but occasionally it may cause exactly the symptom complex of lateral sclerosis, when the lesions are almost confined to the spinal cord. In such cases the differential diagnosis may be impossible.

*Hysteria* may cause a spastic paraplegia, but careful study will usually reveal some evidence of this neurosis. There may be some disturbance of sensation in segmental forms, some ocular signs; or the symptom complex may not be typically that of lateral sclerosis. An ankle clonus, if it occur, is seldom exactly the same as that of organic disease, although it may be found in grave hysteria. This statement is disputed by certain writers. If hysterical the gait will usually afford some clue to the neurosis, and the symptoms may disappear under suggestive treatment.

*Caries of the spine* may occur without kyphosis and without other symptoms than those of implication of the lateral columns by compression. The diagnosis, while not usually difficult, may be impossible.

*Myelitis* may be very hard to distinguish and the clinical picture may be exactly that of lateral sclerosis. It is often syphilitic in character, and examination of the spinal fluid is important.

*Secondary degeneration* from cerebral lesions is usually distinguishable by the presence of cerebral symptoms and the much greater implication of one side of the body. The onset is likely to be acute, with first one side affected in one attack and later the other side in a second attack. The paralysis is more intense, as in lateral sclerosis the spasticity is usually more pronounced than the weakness.

*Combined sclerosis* usually will cause at least slight indication of involvement of the posterior columns; there may be some indication of ataxia.

*Spastic spinal paralysis* must also be distinguished from congenital arrest in the development of the pyramidal tracts or degeneration of these tracts from injury of the brain, with epilepsy, mental defect, etc. Similar cases may be caused by premature birth, intra-uterine cerebral disease, hydrocephalus, etc. The upper limbs may be more affected than they are in the typical spastic spinal paralysis, or cranial nerve implication may be detected.

**Treatment.**—Division of a certain number of the posterior lumbar roots may prove to be of benefit in lessening the spasticity, as may also injection of the nerves supplying the more spastic muscles with alcohol

(Schwab and Allison). These forms of treatment are too recent to permit a judgment as to their value. It is possible that massage and electricity directed only to the less spastic muscles may bring about a more even degree of tonicity in all the muscles. Special forms of movements may be of service.

### HEREDITARY SPASTIC SPINAL PARALYSIS.

**Etiology.**—Syphilis, trauma, etc., seem to have no strictly causal relation, although trauma may act as an exciting cause in persons predisposed. The second of Newmark's cases with necropsy seems to show that agenesis of the pyramidal tracts may exist, so that these tracts from the beginning are imperfectly developed, and if slight degeneration occur it would be likely to produce pronounced symptoms. This hypothesis would explain those mild cases where the only sign of the disease is exaggeration of the tendon reflexes, as in two members of Newmark's O'Connor family. There is much to support the view that the spastic spinal paralysis of family type is an abiotrophy, especially the commencement in several members of the same family at nearly the same age. The motor fibres have limited potentiality, and sooner or later degenerate.

Strümpell thinks the male sex is more predisposed, and that when the males are affected in a family the females always escape, but this latter statement may be challenged. The difference between the sexes as regards frequency is not very striking. The disease is both familial and hereditary. Several observations show, especially Bernhardt's, that in the same family other nervous diseases occur in addition to spastic paralysis. The heredity is not always in exactly the same type, some neurosis or some organic nervous disease of different character may have been present in an ancestor, and yet the cases in which exactly the same form of paralysis has been observed in parent and child are sufficiently numerous to show that the heredity may be direct.

**Pathology.**—The findings are degeneration of the pyramidal tracts extending throughout the cord, most intense in the lumbar and lower thoracic regions, and becoming indistinct in the upper thoracic region (Newmark), or extending in lessened intensity to the pyramidal decussation (Strümpell). The degeneration of the crossed pyramidal tracts in a mild case may be found only in the lower lumbar region, and may be very slight even there, with a little greater involvement of one side of the cord than of the other (Newmark). The direct pyramidal tracts were probably degenerated in one of Strümpell's cases (Gaum).

The direct cerebellar tracts may be degenerated, but not intensely (Strümpell), or may be intact (Newmark). This degeneration may be followed above the pyramidal decussation into the medulla oblongata. The columns of Goll are degenerated, and more in the upper regions of the spinal cord than in the lower. Whereas, Strümpell has found the degeneration of the columns of Goll less intense than that of the crossed pyramidal tracts, Newmark has observed the reverse. The degeneration has been traced to the nuclei of these columns in the medulla oblongata. Newmark found in one of his two cases the net-like condition in the

posterior columns, due to degeneration and dropping out of fibres, such as is seen in cases of severe anemia, and he suggests the possibility of an exogenous factor in addition to the endogenous factor.

Strümpell has maintained that the degeneration of Goll's columns is secondary. If these columns degenerate first and the pyramidal tracts much later, real spasticity could not occur, as shown in Friedreich's ataxia, but Newmark believes, and apparently with reason, that his own second case with necropsy proves that a previous degeneration of Goll's columns does not prevent the spasticity resulting from the degeneration of the pyramidal tracts, as the degeneration of these tracts was only in the lower lumbar region and very slight, while that of Goll's columns was pronounced. Although the degeneration of the last-mentioned columns seems to have occurred in every case with necropsy, in none has it produced recognizable symptoms. The lesions are those of a combined sclerosis. The anterior and posterior roots, and the nerve cells of the anterior horns have not been found affected.

**Symptoms.**—Spastic paraplegia occurs in children or adults of the same family and may be observed in several generations of the same family, sometimes commencing at an earlier age in the later generations. The symptoms, at first with little or no weakness, are spasticity, exaggerated reflexes, usually only in the lower limbs, Babinski's reflex, and paralysis as a late manifestation. Contractures of the feet may be very pronounced, but sensory symptoms are not present; although in Strümpell's case Polster tactile sensation was a trifle less acute in the legs than in the thighs, and temperature sensation was not entirely normal in the legs, although it was so in the thighs. This disturbance could not be attributed to vasomotor changes, as these as well as trophic symptoms were absent. Newmark would not exclude cases in which optic atrophy, feeble mentality, muscular atrophy, bulbar symptoms, bone lesions, and cataract occur. Such cases may not be pure examples of spastic paraplegia, but they are manifestations of abiotrophy. The symptoms usually developed nearly at the same period in the same family, but not always, and the periods vary in different families. In some families the disease first shows itself in the first years of life; in others, in early adult age; in others, still later. In one family Newmark found the ages at which the symptoms began fourteen, seven, nine, sixteen, eight, and eight years respectively. Krafft-Ebing observed the commencement at about twenty-seven, twenty-four, and twenty-two years respectively in members of the same family.

The symptoms may reach their height within a short time, especially after fever or injury, and remain at a certain stage a long time or permanently. In mild cases some improvement is possible. In some cases slow progression begins again after a period of arrest; in others the symptoms progress slowly from the time they first become manifest.

Cases with mental symptoms, implication of cranial nerves, unless possibly strabismus, may be regarded as unimportant, history of traumatism at birth, symptoms existing from birth, epileptic attacks, disturbances of sensation or muscular atrophy, can hardly be regarded as typical of the spinal form of spastic family paralysis. The family form



of spastic paraplegia, probably being the result of degeneration of the distal portions of the pyramidal tracts from some imperfection of development, differs from the cases of Little's disease resulting from premature birth, in that in the latter the pyramidal tracts are arrested in their development, but are capable of further development even many years after birth, and probably do not degenerate. In the form occurring in childhood, the pyramidal tracts probably degenerate at their distal ends early, while in Strümpell's type the resistance of these tracts seems to be greater, and disturbance of function does not occur until adult life.

In some families the cerebral type seems to occur in certain members, and the spinal type in others (Bernhardt, Pribram, Melotti, and Cantal-emessa). This would seem to indicate that there may be a close relation between these two types. In a family studied by the writer, a father and his second son showed clearly the typical symptoms of the disease, and a younger son showed indications of it. Only the lower limbs were affected. The disease seemed in the latest generation to be commencing at an earlier period. It has been almost as common in this family in the female sex as in the male. In late years the disease is said to have commenced before or about the fifth year of life, and it has always presented the same symptoms. A young member of the family manifested the first symptoms about the age of eighteen months. The disease in this family was traced back five generations. This family is doubtless the same as that reported at an earlier date by Bayley, although the writer was not aware of this fact at the time of his report.

Dejerine and Thomas say the affection extends later to the upper limbs and eyes and to the muscles of speech, and yet in most cases these parts, except for exaggeration of the tendon reflexes, escape.

In late stages the lower limbs may be almost completely paralyzed; Strümpell says it is characteristic of the hereditary spastic paralysis to have great exaggeration of tendon reflexes precede many years true paralysis. It is uncertain whether reflex exaggeration and weakness result from disease of different fibres or from different degrees of degeneration of the same fibres. Strümpell has not seen involvement of the upper limbs beyond exaggerated reflexes. The condition must be distinguished from the spastic cerebral paralysis of childhood, not always with defect of mentality. This is an agnesia and may be familial in type and has an etiological relation to the spinal form.

**Diagnosis.**—This must be made from cerebral paralysis, Little's disease, myelitis, Pott's disease, lateral sclerosis, family form of disseminated sclerosis, Friedreich's ataxia, and cerebellar hereditary ataxia. The difficulties are those mentioned for primary lateral sclerosis. Strümpell acknowledges that he, like Charcot, diagnosed primary sclerosis in a case in which the necropsy showed multiple sclerosis.

In some cases bulbar and cerebral disturbances existed in addition to the spastic paraplegia (Bernhardt), suggesting multiple sclerosis; in others premature birth, strabismus, slow acquirement of speech, feeble intelligence suggested cerebral origin. Family spastic paraplegia may be cerebral when associated with such symptoms, and Dejerine and Thomas would regard it merely as a symptom complex common to family

disease of different character and situation. It seems that cases in which the upper limbs are contracted, intention tremor, scanning speech, nystagmus, atrophy of the optic disks, strabismus, etc., are present, should be accepted with reservation. They do not belong to the typical form and may be examples of multiple sclerosis.

### UNILATERAL ASCENDING AND DESCENDING PARALYSIS.

Attention was first called to these forms of paralysis by Charles K. Mills. In many cases it is a symptom complex instead of a well-defined disease, just as bilateral spastic paraplegia is often a symptom complex, but may be the clinical expression of primary degeneration of the pyramidal tracts. The question that now interests us is, Does unilateral primary degeneration of the pyramidal tract occur? Some years ago the writer searched for all the cases of the unilateral form of amyotrophic lateral sclerosis in the literature, and found ten; to these should be added a case reported by Potts, one reported by the writer, and one by Mills. The unilateral type of amyotrophic lateral sclerosis is only of short duration, and bilateral implication of the spinal cord occurs within a short time. The 13 cases are those of Dejerine, Pick, Vierordt, Leyden, Lennmalm, Mott, Senator, Probst (3, possibly 4), Potts, Spiller, and Mills. Of these, only 5 were of the ascending type, the weakness commencing in one lower limb and later extending to the upper limb of the same side. These were the cases of Vierordt, Mott, Probst, Potts, and Spiller. The symptoms may begin with bulbar palsy (Dejerine, Leyden, Lennmalm, Probst (2 cases) ). The upper limb of one side may be affected before the lower (Pick, Leyden, Probst). In some of the cases the limbs of one side seem to have been affected simultaneously or nearly so (Dejerine, Lennmalm, Senator, Probst (2 cases, Mills). In those cases in which a necropsy was obtained the pyramidal tract of each side was degenerated. It is yet to be shown whether amyotrophic lateral sclerosis may ever remain confined to one side; at present it seems improbable.

A case of primary lateral sclerosis, reported by Mills and Spiller, would seem to indicate that the unilateral form of the disease without implication of the nerve cells of the anterior horns is possible. The patient, a male, aged sixty years, developed gradually hemiplegia on the right side, the lower extremity being more markedly, and for this reason probably earlier, affected than the upper; the case, therefore, at first belonged to the clinical type of unilateral progressive ascending paralysis. After several years the left lower extremity also became paralyzed, but not to the same extent as the right. The reflexes were all markedly exaggerated, the Babinski response being present. Sensory symptoms were absent. The lesions were primary degeneration of the motor tracts, much greater and of longer duration in the right crossed and left direct pyramidal tracts, with integrity of the nerve cells of the anterior horns.

It seems positive that unilateral ascending or descending paralysis may be caused by different conditions. It may, in the opinion of Mills, be the expression of a gradually developing cerebral lesion such as softening or tumor, of disseminated sclerosis, of a developing posterolateral

PLATE I



Progressive Neural Muscular Atrophy.

Atrophy of the hands and forearms, with escape of the upper arms in the writer's case, lasting about forty-five years.



PLATE II



Atrophy and Contracture of the Feet and Legs.

With escape of the thighs, in the case represented in Plate I.



sclerosis, of spinal syphilis, of tumor of the cord or of other form of compression of the cord, of paralysis agitans, of hysteria. The type has varied in different cases even when the degeneration has been believed to be confined to the pyramidal tract. In some spasticity has been present from an early period, in others it has been absent or slightly marked. The tendon reflexes have usually been exaggerated, but in a few instances have been diminished or slight. The wasting of the muscles has varied in degree. Optic atrophy and vesical disturbance have been observed. The explanation of these differences is to be sought chiefly in the greater or less implication of the cells of the anterior horns of the spinal cord.

Pathological evidence as yet does not warrant the assumption that degeneration of the crossed pyramidal tract may begin in the lower part of this tract, or in the upper part, gradually extend, so as to implicate the motor fibres for the other limb of the same side, and remain confined to one pyramidal tract.

#### **PROGRESSIVE NEURAL (NEUROTIC) MUSCULAR ATROPHY (PROGRESSIVE MUSCULAR ATROPHY OF THE CHARCOT-MARIE-TOOTH TYPE).**

Charcot and Marie, and Tooth described this form of atrophy independently in the same year (1886). The subject soon received attention from Joffroy, Hoffmann, B. Sachs in this country, and from many others, but evidently much has been included that does not properly belong here.

**Etiology.**—No satisfactory cause has been found, but the fact that it is likely to occur both as an hereditary and as a family disease shows that some defect in development is probably responsible for it. It is more likely to occur in males (five times in males to once in females, according to Sainton), but no explanation for this can be found. The disease may show a similar heredity, *i. e.*, father and son may both be afflicted in the same way, and this is true also of the spastic family paralysis. It has usually begun before the age of twenty-two years.

**Pathology.**—The lesions are sclerosis of the posterior columns of the cord, slight degeneration of both pyramidal tracts (Sainton), with integrity of the anterolateral columns in some instances (Marinesco), alteration of the columns of Clarke, atrophy of the cells of the anterior horns of the cord, alteration of the peripheral nerves, which may be slight, and of the intramuscular branches, atrophy of muscle fibres, and chronic meningitis (Dejerine and Armand-Delille). The nerve trunks, the cutaneous sensory nerves, and the anterior and posterior nerve roots, with slight exception, may be normal.

**Symptoms.**—The characteristic features of this form of atrophy as given in the first description by Charcot and Marie are: Progressive muscular atrophy implicating first the feet and legs, and not appearing in the upper limbs (hands and, later, forearms) until several years later, the progression of the atrophy being slow. Relative integrity of the muscles of the limbs near the trunk, or at least much longer preservation of these than of the muscles of the distal ends of the limbs. Integrity of the

muscles of the trunk, shoulders, and face. Fibrillary contractions in the atrophying muscles. Vasomotor disturbances in the portions of the limbs atrophied. No pronounced contractions of tendons in the atrophied limbs. Sensation usually intact, but sometimes affected. Cramps frequent. Reaction of degeneration in the atrophying muscles. Commencement of the affection usually in childhood, the disease often found in several brothers and sisters, and sometimes in the previous generations.

Charcot and Marie, in their first publication, acknowledged the possibility of implication of the muscles of the limbs near the trunk, as they speak of relative integrity of these muscles. They also stated that the muscles of the thigh seem to preserve their power and volume during a certain period, but that this integrity often is not real. The vastus internus is the first of the thigh muscles involved. Undoubtedly they emphasized the earlier and greater implication of the muscles at the distal ends of the limbs as the most characteristic feature of the disease. Cases absolutely typical in other respects have existed with grave contractures of the tendons of the feet.

The implication of the distal portions of the limbs, with complete or nearly complete integrity of the proximal parts, is the essential feature of the disease. So soon as we permit cases with intense atrophy of the proximal parts of the limbs, and especially with atrophy of the muscles of the trunk, to be classified under this designation, the diagnosis is probably incorrect. Typical cases with necropsy reported in the literature are those of Marinesco, Sainton, and Dejerine and Armand-Delille. The cases of Dubreuilh and Siemerling are questionable.

The tendon *reflexes* are usually lost, but at least one case, although without necropsy, is reported in which they were exaggerated (Dercum). This is so extraordinary, and so in contradiction to what we know of the pathology of the affection, that when it occurs we must think of a complicating lesion, such as pronounced degeneration of the lateral columns.

Although the atrophy begins so frequently in the distal parts of the lower limbs that the type has been designated by Tooth as the peroneal, it may begin in the upper limbs, but its commencement is always in the peripheral parts of the limbs, and the progress of the disease is slow. A few cases have been observed in which the atrophy began in the distal parts of the upper and lower limbs nearly at the same time.

*Contractures* (talipes equino-varus) are not uncommon in the muscles controlling the movements of the feet, and often they impede the gait very greatly. The atrophy of the legs below the knees may be excessive, and when this stage has been reached the lower parts of the thighs will usually be found comparatively less well developed than the upper parts. All movements of the toes or of the foot at the ankle may be impossible, both because of the atrophy and because of the contractures. The hands may present the condition seen in progressive spinal muscular atrophy, with wasting of the interosseous spaces and contracture, the so-called griffon's claw. The lower part of the forearms may be intensely atrophied, but the upper part is usually in a more nearly normal condition. Fibrillary contractions occur in the wasting muscles, just as they occur in all muscular atrophy when the cells of the anterior horns are diseased.



Vasomotor disturbances are common. The wasted limbs feel colder to the touch and appear more or less cyanotic. Sensation may be disturbed in the peripheral parts of the limbs, or pain may be felt, but, as a rule, the sensory symptoms are not nearly so pronounced as the motor.

The electrical reactions are often altered, and this alteration may vary from diminution in the intensity of the electrical response to reaction of degeneration or even complete loss of the electrical response. Diminution in the faradic response of nerves in regions where muscular atrophy has not been as yet detected has been emphasized especially by Hoffmann.

Mental symptoms, implication of the sphincters, pupillary phenomena, are not part of the symptom complex.

The disease is of slow development, and after reaching its height the patient may live many years without additional symptoms. Thus, in a typical case reported by the writer, the symptoms are said to have been present about forty-five years, and the patient's condition has not changed during the many years he has been under observation. There is not likely to be any tendency toward improvement.

The ocular manifestations have been studied by W. Krauss. They are not frequent, or at least little has been described concerning them. Krauss has found pupillary changes mentioned four times, which, however, are inaccurate, and in 3 or 5 cases atrophy of the optic nerve. He concludes that the only reliable ocular manifestation is optic atrophy.

**Diagnosis.**—When the symptom complex is typical the diagnosis is easy. The hereditary or family tendency, the youthful age at which the symptoms commence, the slow progress, the limitation of the atrophy to the peripheral parts of the limbs, the loss of tendon reflexes, the motor disturbances much greater than the sensory—all these make a definite clinical picture. The diagnosis will not be difficult if we refrain from including cases with atypical features.

*Progressive muscular dystrophy* is most liable to cause mistakes, and a case with necropsy reported by the writer shows the difficulty in diagnosing between these two diseases. The atrophy began in the lower parts of the lower limbs, and was associated with slight reaction of degeneration in these parts and talipes equino-varus on each side. These and the symmetry of the atrophy suggested the neurotic form, but the extension of the atrophy to the proximal parts of the limbs and the muscles of the trunk was unlike the typical form of the neurotic muscular atrophy. The microscopic examination showed that the nervous system was normal, but the muscles were intensely degenerated.

*Anterior poliomyelitis* with paralysis confined to the distal parts of the lower limbs may occur and cause some confusion, but when a history of the onset is obtainable the diagnosis is easy.

*Progressive spinal muscular atrophy* or chronic poliomyelitis may resemble the Charcot-Marie-Tooth form, but these diseases are neither hereditary nor familial, do not occur at so early an age, nor does the atrophy remain confined to the distal ends of the limbs.

*Multiple neuritis* usually does not implicate the peripheral portions of the limbs, and the sensory disturbances are pronounced. Motor neuritis, with little or no sensory involvement, does occur, and the

Charcot-Marie type is regarded by some as a motor neuritis in which the peripheral ends of the longest nerves are affected; there is much to be said in favor of this view, although the spinal cord is also implicated.

### INTERSTITIAL HYPERTROPHIC PROGRESSIVE NEURITIS OF CHILDHOOD.

This disease was first observed by Dejerine and Sottas, and was reported by them in 1893. They had two cases occurring in brother and sister, and necropsy was obtained in both. They referred to a case of Gombault and Mallet as belonging to this type, and this case was the first published, although it was considered by the authors as a form of tabes. Syphilis may have some causal relation; at least, in the second case of Dejerine's a history of syphilitic infection at the age of twenty-four years was obtained. Alcoholism and poor mental development were present in this second case.

Marinesco regarded the disease as a variety of the neurotic muscular atrophy, and this view Brasch takes in the report of two clinical cases, although it has been most vigorously combated by Dejerine, who mentions that in the Charcot-Marie-Tooth form incoördination of movements, marked disturbance of sensation, Argyll-Robertson pupils, kyphoscoliosis, and hypertrophy of nerves do not occur, no matter how long the affection may have lasted. The duration of the affection has nothing to do with the hypertrophy of the nerves, according to Dejerine.

Marie presented at the Neurological Society of Paris, in 1906, two cases occurring in a family of seven children, all similarly affected. He regarded the disease as a special form of the interstitial neuritis, although his cases differed in certain respects from the cases of Dejerine and Sottas. Dejerine accepts Marie's cases. They presented the same deformity of the feet, kyphoscoliosis, loss of reflexes, diminution of sensation, disturbance of gait and station, muscular atrophy, and considerable hypertrophy of the nerve trunks. Marie believed the muscular atrophy was different in his cases, inasmuch as it affected only the muscles of the feet and legs, while in the cases of Dejerine the atrophy was general. Marie did not find the Argyll-Robertson sign, but the light reflex was diminished; he also noted absence of disturbance of the sphincters and of the sexual functions.

**Pathology.**—The lesions of the spinal cord are only such as result from degeneration of the posterior roots, and are like those of tabes. The lesions of the nerves are very characteristic, and are parenchymatous and interstitial. The nerve fibres may be altered to such a degree as to leave only empty neurilemma sheaths, but only a certain number are so intensely degenerated. The connective tissue is greatly thickened, and the alteration begins in the periphery and extends to the nerve trunks and nerve roots. The lesions are more pronounced in the nerves and roots of the lower limbs than in those of the trunk and upper limbs, and still more so than in the bulbar nerves; but only the optic and olfactory nerves escape. Such hypertrophy occurs in no other form of neuritis.

**Symptoms.**—These may be said to be ataxia of the four limbs with muscular atrophy, marked disturbance of sensation with retardation

of sensation, fulgurant pains, nystagmus, myosis, inequality of the pupils, Argyll-Robertson's sign, kyphoscoliosis, marked hypertrophy and hardness of all nerve trunks accessible to palpation, *i. e.*, the symptoms of tabes associated with general muscular atrophy, kyphoscoliosis, and hypertrophy of nerves.

Muscular *atrophy* may vary in different cases; in some it may be very intense, and predominate in the distal parts of the limbs, diminishing toward the trunk. The muscles of the face may be paralyzed and atrophied, and the facies may resemble that of myopathy, with the prominent lips and protrusion of the upper lip, the lips of the ant-eater (*tapir*), with the transverse laugh (*rire en travers*), so that the corners of the mouth are not drawn upward, and with difficulty to pout the lips or whistle. The distribution of the upper branch of the facial so far has escaped. The feet are always in the varus position, the nails are deformed, the first phalanx of the toes is dorsally flexed, and the second and third are plantary flexed upon the first, forming a right angle with it.

The disturbance of function in the lower limbs is the result of atrophy and ataxia. The gait is steppage, but not like that resulting from paralysis of the extensors of the toes and of the anterior tibial muscle. The steppage is abrupt, the legs are raised and thrown outward, the feet falling quickly upon the ground. A cane is necessary in walking, and the head is held forward, and the steps are carefully watched. Turning around is done slowly, and with caution. Ataxia is pronounced when the feet are together and is pronounced in the upper limbs. The muscles of the larynx may be paralyzed, as in tabes, but, unlike tabes, the sphincters and genital powers are always intact.

The nerves may be double the normal size, and the hypertrophy is uniform without nodes of swelling. Pressure on these enlarged nerve trunks does not cause pain; on the contrary, the nerves are analgesic to pressure and electrical irritation.

There can be no doubt that the interstitial hypertrophic neuritis of childhood resembles the neurotic muscular atrophy in certain of its symptoms, but in others it resembles tabes dorsalis; and yet it is impossible to regard it as an association of the two diseases in the same person. There is evidently a strong family tendency to the affection.

The *treatment* is unsatisfactory. The pains may be relieved by analgesics, and the ataxia lessened by coördinated movements. The treatment is much like that of tabes.

### PROGRESSIVE BULBAR PALSY (GLOSSO-LABIO-LARYNGEAL PARALYSIS).

**Etiology.**—No cause is known, but it is probably an abiotrophy. The motor cells of the medulla oblongata and pons have a vitality sufficient to enable the functions of life to be properly performed until about middle age, or later, and then they begin to degenerate and finally disappear. No reason can be given for the escape of the sensory nuclei in the same regions. Syphilis does not seem to play a rôle, although it is possible that lead may do so. Infectious diseases may cause acute

bulbar palsy by producing a bulbar encephalitis, but it is questionable whether they ever give origin to chronic bulbar palsy. Bulbar palsy may develop as a result of electricity. It is exceedingly rare in early life, but the occasional occurrence at that period and in several members of the same family can be explained only by a congenital tendency.

Most authors state that males are more frequently affected. In the writer's experience females have been as often subjects of the disease as males. The patient is usually in or past middle life before the onset.

**Pathology.**—The nuclei of the hypoglossal nerves are usually intensely altered, so much so that nerve cells in these nuclei may have disappeared; in other cases they are few in numbers and much atrophied, with loss of dendritic processes. The intramedullary fibres of the hypoglossal nerves become atrophied and may also disappear. The alteration of the nucleus ambiguus may be very pronounced, but great care is requisite in forming an opinion concerning its condition. The posterior nuclei of the pneumogastric nerves in a case examined by the writer were so intensely pigmented that they appeared degenerated. The nuclei of the facial nerves and more rarely the motor nuclei of the trigeminal nerves are degenerated. The nerves whose function is interfered with are the hypoglossal, facial, pneumogastric, glossopharyngeal, vago-accessorial, and motor trigeminal. Hemorrhages or round-cell infiltration are not found in true progressive bulbar palsy. The process is gradual, and atrophy and degeneration of the motor nuclei and nerves of the medulla oblongata and pons occur.

Raymond believed that progressive bulbar palsy is almost always a part of amyotrophic lateral sclerosis, although it may occasionally exist alone. Leyden did not admit the latter as a possibility, and Dejerine mentions that in 1883 he demonstrated that the lesion is not confined to the bulbar nuclei, and that the pyramids are distinctly affected. The bulbar paralysis of Duchenne is a pyramidal amyotrophic sclerosis descending in type. With the exception of Reinhold's case (1890), which is not entirely satisfactory, no necropsy of a case of labio-glosso-laryngeal paralysis exists with integrity of the pyramidal tract. Remak's case (1892), quoted by some writers as one with integrity of these tracts, is not one of glosso-labio-laryngeal paralysis of Duchenne, as the superior facial branch was as much implicated as the lower, and the elevators of the eyelids were paralyzed. These symptoms belong to polioencephalitis, but not to Duchenne's bulbar palsy. The latter may occur in two forms, alone or in association with the symptoms of amyotrophic lateral sclerosis. The lesions of the bulbar nuclei are the same in either case, and the pyramidal tract is involved, as demonstrated by exaggeration of the tendon reflexes when the picture is typically that of Duchenne's bulbar palsy.

Chronic poliomyelitis, it is said, never terminates by bulbar palsy. Notwithstanding this statement, reasoning by analogy would lead us to believe that uncomplicated bulbar palsy might occur. Dejerine himself has reported two cases in which the nerve cells of the anterior horns of the spinal cord were degenerated without implication of the motor tracts, and if this may occur in the spinal cord it is hard to under-

stand why it may not occur in the medulla oblongata and pons. In several instances the writer has found bulbar palsy in association with amyotrophic lateral sclerosis, but never with integrity of the motor tracts.

The muscles of the tongue are intensely atrophied, and if examined by the method of Marchi may show the presence of fat drops within the muscle fibres. The muscles of the lips and larynx also may be atrophied.

**Symptoms.**—The clinical picture is most distressing. The patient has difficulty in speaking, progressing to loss of speech; dribbling of saliva, disturbance in swallowing, so that fluids return through the nose, and with much coughing, from the entrance of some of the fluid into the larynx. The ability to pucker the lips, to whistle, or blow out a lighted match is lost, and with all this, intelligence is preserved, and the unfortunate individual has no dulling of mentality. The disease is remarkably symmetrical in its development.

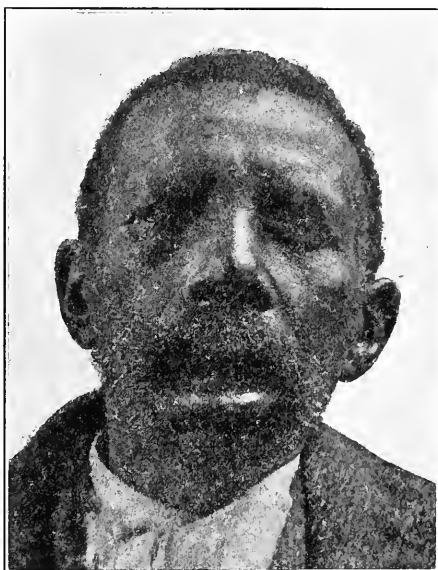
**Speech.**—Difficulty in pronouncing certain letters—d, g, k, l, r, s, t—is at first noticed; the words are slurred, and gradually speech becomes so indistinct that those who have known the patient many years and are familiar with his voice fail to understand what he says. Beevor observed one case in which the disturbance was observed in singing before it was perceptible in conversation. The defect at first may be noticed only after the patient is a little fatigued in speaking. The voice becomes nasal because of paralysis of the soft palate, and when the larynx is paralyzed the defect of speech has reached its height. The

tongue is atrophic, and may be so intensely affected that it cannot be protruded beyond the lips, is full of wrinkles, and shows an intense degree of fibrillary tremor. The reaction of degeneration may be obtained. The weakness of the tongue interferes with mastication of food, as the food cannot be controlled by the action of this organ, and pushed into the pharynx. The atrophy of the lips may be masked by the deposit of fat in the tissue.

The muscles of mastication are less often affected, but may be weak, so that mastication of food becomes difficult. The atrophy and weakness in the distribution of the facial nerve is limited almost to the muscles of the mouth and chin, so that the upper part of the face is not involved, except in the infantile form of bulbar palsy.

Paralysis of the pharynx makes deglutition impossible when the

FIG. 4



Bulbar palsy.

paralysis is complete, but the weakness develops gradually. Liquids then not only may ascend into the nasal passages, but solids may pass into the larynx and thus cause bronchitis or aspiration pneumonia. The reflexes of the soft palate and pharynx are much impaired. The dribbling of saliva may be so great that patients are obliged to wear bibs.

The upper part of the facial distribution escapes in progressive bulbar palsy, and this peculiar limitation of the paralysis has led to the conclusion by some neurologists that the centre for the upper distribution probably has a situation remote from that of the facial nucleus, and possibly is in the oculomotor nucleus.

The tendon reflexes of the upper and lower limbs are usually exaggerated, and there may be a pronounced jaw-jerk. Beevor described a lower jaw clonus.

The disease is essentially motor in its manifestations, and sensory disorder or enfeeblement of the intellect does not occur. Intelligence is preserved until near death, but the patient is often emotional, and for a long time maintains a hopeful attitude.

The heart may become affected, the pulse rapid, weak, and irregular, and the muscles of respiration impaired. When these signs develop, death cannot be delayed long. The end may be from inanition.

The gait is often peculiar. The steps may be short and slow, but this form of progression is more common in pseudo-bulbar palsy.

In some instances ocular palsies have been associated with bulbar palsy, but they are to be regarded more as epiphenomena, and not as an essential part of the disease.

**Diagnosis.**—The greatest difficulty may be in regard to *pseudo-bulbar palsy*, and yet in typical cases this is not great. The writer has seen pseudo-bulbar palsy, however, in which the only distinguishing features were the absence of atrophy, of fibrillary tremors, and of reaction of degeneration in the tongue. Pseudo-bulbar palsy is caused by multiple lesions in the cerebrum, implicating the nerve fibres innervating the motor nuclei of the medulla oblongata and pons. As the lesions are supranuclear, atrophy, fibrillary tremors and reaction of degeneration do not occur.

*Multiple neuritis* confined to the cranial nerves may occur, but it is seldom that the typical picture of bulbar palsy will be presented; often nerves other than the cranial are affected, and sometimes sensory as well as motor nerves are implicated, and the development is usually more rapid in multiple neuritis. Following diphtheria, the soft palate may be paralyzed as well as the sphincters of the eyes and mouth, the frontalis may be weak, and the muscles of expression about the mouth may escape, although the tongue may be affected, and there may be difficulty of deglutition.

*Myasthenia Gravis Pseudo-paralytica.*—The resemblance of this disease to progressive bulbar palsy may be very striking, but the rapid exhaustion after slight exertion, the myasthenic reaction to electricity, the implication of ocular nerves and of the limbs, the absence of atrophy, at least in most cases, make the diagnosis possible.

*Tumor, hemorrhage, softening, or encephalitis* of the pons and medulla oblongata does not cause symmetrical lesions, does not develop in the same way, and implicates sensory fibres as well as motor.

The *bulbar type of muscular dystrophy*, as described by Hoffmann, is so rare that it may almost be ignored. The presence of other signs of muscular dystrophy should make the diagnosis easy.

*Bulbar palsy* may be a part of *tabes dorsalis*, but in all the cases except those of Charcot and Pfeifer the tabetic symptoms occurred first. The symptom complex differs from that of progressive bulbar palsy in the implication of sensory cranial nerves, so that the diagnosis of *tabes* associated with progressive bulbar palsy could hardly be maintained. Optic atrophy, disturbance of deglutition, reflex rigidity of the iris, ocular muscle palsies, and sensory disturbances in the distribution of the trigeminal nerve were present in all the cases; and weakness of the muscles of mastication in all the cases except one. Increased flow of saliva and facial paresis were observed in most of the cases, and only the distribution of the lowest branch of the facial was affected. Disturbances of coördination in the muscles of the face, associated movements, and involuntary movements were present in some of the cases. Atrophy of the tongue was seen in only about half the cases, and was not complete in any case, this being unlike the condition in progressive bulbar palsy.

A *siphilitic* infiltration of the medulla may give symptoms like those of true bulbar palsy.

The *bulbar palsy of childhood* has been observed only a few times. In the two young brothers studied by Londe the symptoms began by bilateral paralysis of the upper branch of the facial nerve, with greater intensity of one side. Winking was infrequent, and the eyes could not be opened quickly. In one case slight ptosis was present. There was at first no paralysis of the muscles of the eyeballs. In the beginning of the disorder the facies resembled that of myopathy of the Landouzy-Dejerine type. It differed from the facies of labio-glosso-laryngeal paralysis, as in the latter the lower part of the face is affected, and in Londe's cases the upper part of the face at first alone was affected. Gradually in Londe's cases the double superior facial palsy extended, and the entire facial distribution became implicated as well as the oculomotor, the hypoglossal, the spinal accessory, the trigeminal, and the pneumogastric nerves. The lips became inactive and flaccid, without being protruded. The face became an immobile mask, and the smile was impossible. Fibrillary tremors occurred in the chin. The tongue was paretic and atrophied. The muscles of mastication were affected in one case. Paralysis of the abductors of the larynx occurred in one of the brothers.

**Treatment.**—Little can be done to arrest the development of progressive bulbar palsy, and nothing to stop it. The feeding with liquid or semi-liquid food is important, and tonics, especially strychnine, may be used.

### PROGRESSIVE MUSCULAR DYSTROPHY (PRIMARY MYOPATHY).

**Etiology.**—Like in the other affections considered in this chapter, abiotrophy is the chief cause of this disease. Here, however, the muscu-

lar system instead of the central nervous system is affected, and its vitality may be so slight that wasting begins in some cases soon after birth. It is not improbable that some acute infectious disease may be the exciting cause, but the predisposition already exists. The resistance of the muscles varies in different cases, and, therefore, the time of commencement of the disease varies. Heredity and familial occurrence demonstrate clearly the congenital weakness of the tissues, and the mother seems to be more liable to transmit the disease than the father. There are many cases in which no heredity or familial tendency can be traced. A few cases have been observed in which the disease seemed to develop after typhoid fever (Friedländer, Jossierand, Guillain).

The disease first makes its appearance in early childhood, but cases are on record in which it began in middle life or even later. Erb believed that functional alteration of the nerve cells, undetectable by any means at our command, may cause the alteration of the muscle fibres, so that in this way muscular dystrophy may be of nervous origin.

Convulsions, as well as diabetes insipidus, nystagmus, hemiparesis, hysteria, chorea, and psychoses have been observed with muscular dystrophy. Such combinations, in Erb's opinion, are too frequent to be without influence on our judgment regarding the central origin of the dystrophy. Hypertrophied fibres also have been seen in poliomyelitis (Müller, Dejerine), and Hitzig and Kawka have found the same changes in the muscles in this disease as in muscular dystrophy.

Direct heredity in muscular atrophy can no longer, according to Hoffmann, possess the diagnostic value which it was once supposed to have, although it must be conceded that it is more important in the myopathic and neurotic forms (Charcot-Marie-Tooth type) than in the myelopathic variety. At first the Leyden-Möbius type was regarded as the only form of muscular dystrophy hereditary in origin. Onuf has found six cases of epilepsy in which symptoms of muscular dystrophy were present. The congenital tendency was well shown in a family observed by Bunting, in which three male members were affected at the same age (five years). The boys of this family were susceptible, the girls escaped. It seems extraordinary that sex should influence the onset of this disease, especially at so early an age.

**Pathology.**—The lesions are in most cases purely muscular, the atrophy may be so intense that no muscle fibres remain, in other cases the fibres are smaller in size, and yet the muscle spindles are intact, even in the most extreme atrophy of the surrounding muscle. It is probable that these spindles are organs of sensation and possibly are concerned with the sense of position.

Landouzy and Dejerine believed muscular dystrophy to be a progressive atrophic myopathy resulting from a primary parenchymatous myositis with hypertrophy of certain muscle fibres, terminating in "simple" atrophy. The interstitial myositis is slight, and fatty infiltration may be found in some muscles. The vessels and intramuscular nerves, they stated, had never been found diseased. Some investigators have believed that the disease begins in the interstitial tissue of the muscles, others in the vessels, and still others in the muscle fibres.



The findings are hypertrophy and atrophy of muscle fibres, proliferation of the nuclei, vacuolation, splitting of muscle fibres, hyperplasia of connective tissue and proliferation of its nuclei, thickening of the walls of the vessels and proliferation of their nuclei, accumulation of fat cells.

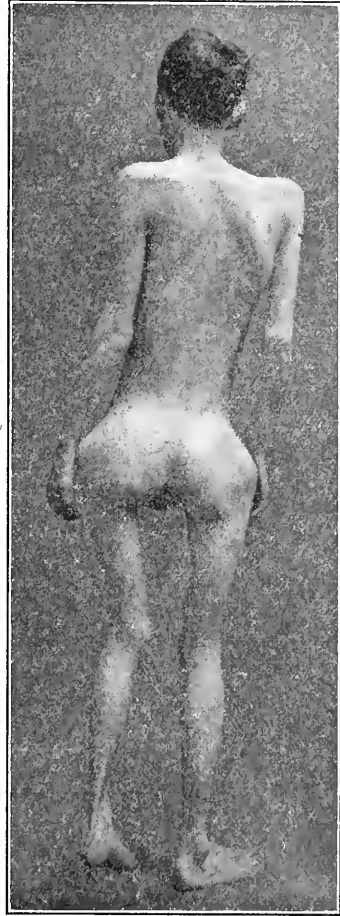
Erb mentions that more or less important changes have been found in the anterior horns of the spinal cord, but that negative findings are the rule, the positive the exception. These cellular changes are important in the understanding of the pathology, as they demonstrate that the most typical clinical manifestations of progressive muscular dystrophy may be with important alteration of the nervous system, although we do not know why this should be; whether it depends on the type, duration, intensity, external causes or complications. The changes in the muscles are primary, those in the connective tissue secondary.

The question as to whether the atrophy of muscular dystrophy differs from that of other processes has interested many, and the question arises whether the histological findings are sufficient to permit a diagnosis of the disease. Erb acknowledged that muscular findings similar to those of dystrophy might occur in other diseases, as they had been seen in spinal disorders, in myositis, near tumors situated in muscles, and in muscles in regeneration after injury.

**Symptoms.**—The characteristic features are: The early age of commencement. The hereditary or familial appearance. The portions of the body affected, the atrophy beginning in the proximal portion of the limbs and trunk. Pseudo-hypertrophy of certain parts of the body, especially of the calves. Quantitative diminution of electrical responses, depending on the degree of atrophy, but reaction of degeneration absent in most cases. Diminution or loss of tendon reflexes. Absence of fibrillary tremors.

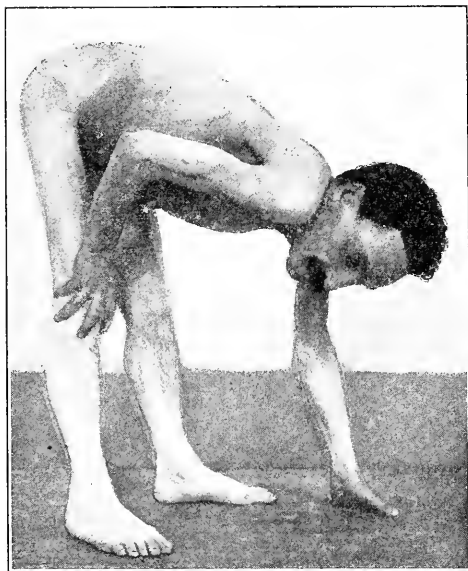
The *atrophy* almost invariably begins in the proximal parts of the body and the muscles of the trunk; in some instances the muscles of the shoulder girdle are affected first, in others those of the lower part of the trunk. The atrophy, however, is not always the first sign, for the muscles of one calf may enlarge in association with a little weakness of the leg, before any atrophy can be detected. In rare instances the atrophy begins in the distal portions of the limbs.

FIG 5



Progressive muscular dystrophy. Intense and general muscular wasting.

FIG. 6



The method of rising from the floor as shown in progressive muscular dystrophy. This is seldom seen so late in life as in the patient represented, inasmuch as the disease begins usually in childhood, and by middle life the disability has become too great.

the forearms and hands are likely to show wasting. In the lower limbs the muscles commonly affected are the gluteal, and the muscles of the anterior and posterior parts of the thighs. The muscles of the face and neck are only occasionally wasted, and when this occurs a distinct type is produced. Usually, in such cases, only the muscles of expression are involved, but the masseters, tongue, and ocular muscles have been found implicated a few times.

In a patient observed by Marie closure of the eyelids was impaired, as in the infantile type, and marked double ptosis existed. The muscles of mastication were affected in the same case. Marie has found a few cases reported in which either ptosis or implication of the muscles of mastication occurred alone in muscular dystrophy, but no case in which both occurred together.

*Weakness* usually precedes atrophy, and at first is confined to certain portions of the body; later the muscles of all the limbs as well as those of the trunk may be greatly wasted. Where two or more members of a family are affected the atrophy may show different types, not commencing in corresponding parts in members of the same family, but occasionally the same type is preserved in a family. Dystrophy of the muscles of the shoulder girdle in father and son was observed by Ogilvie and Easton. The shoulder girdle is frequently implicated, and the muscles affected are the pectorals, supraspinatus, infraspinatus, trapezius, rhomboid, subscapularis, biceps, triceps, brachialis anticus, and supinator longus; and it is not until late that the muscles of

FIG. 7



The transverse laugh (*rire en travers*) seen in facial type of progressive muscular dystrophy.

The *pseudo-hypertrophy* attacks most frequently the muscles of the calves. This is caused by overgrowth of fibrous and fatty connective tissue, although here and there true hypertrophy of certain muscle fibres may be found in the midst of the atrophied muscles, but they do not cause any increase in the bulk of the muscle. In some cases the pseudo-hypertrophy may occur in the upper limbs in the triceps or deltoid muscles. The atrophy is usually seen in groups of muscles functionally associated.

Reactions of degeneration should arouse suspicion that the atrophy may be both myopathic and myelopathic. Certain cases have been reported of the myopathic type in which it has occurred.

Fibrillary tremors do not belong to progressive muscular dystrophy, but they have been observed in some cases. They are of much value in diagnosis when the nerve cells of the spinal cord and bulb are believed to be affected. When they occur in muscular dystrophy they are usually confined to one or a few muscles, and are usually of transitory duration.

The condition of the tendon reflexes depends, in large measure, upon the degree of atrophy. Where the quadriceps femoris is much wasted the patellar reflex must be diminished or lost, but in some cases this reflex may disappear before any diminution in the size of the muscles can be detected. It is presumable that some change has begun in the muscle fibres, making them less capable of responding to stimulation.

Peculiar attitudes may be caused by the wasting of certain groups of muscles. Where the shoulder girdle is attacked the condition of "winged scapula" and "loose shoulders" may be produced. If the patient be lifted by the axillæ the shoulders are carried so far upward as the tissues permit. When the muscles of the back are wasted lordosis may be very pronounced, in rare cases it may be so extreme that the abdomen rests upon the thighs. More rarely lordosis is caused by weakness of the abdominal muscles. A condition occurring in muscular dystrophy, in which the muscles of the neck are affected, has been called by Ballet and Delherm "*facies of the sphinx*." It consists of an enlargement transversely of the base of the neck and flattening in the anteroposterior diameter. It is associated with atrophy of other muscles, and is said to be common in muscular dystrophy.

Contraction of the biceps tendon so that the forearm cannot be extended has been noted especially by Landouzy and Dejerine, but it is not of common occurrence, and probably depends on pronounced muscular atrophy. The flexor muscles on the back of the thighs are sometimes much contracted, and the legs are greatly flexed upon the thighs. Retraction of muscles occurs early or late in the disease, especially in the biceps of the arm, flexors of the knee, and muscles of the calf. The cause is not very evident. Where contracture is the first sign of dystrophy it is probably not produced by retraction of fibrous tissue.

The wasting of the gluteal muscles causes a peculiar elevation of first one hip then of the other in walking, the duck walk (*marche de canard*), as it has been called by the French writers. It is extremely characteristic of progressive muscular dystrophy.

The weakness of the muscles of the back causes a peculiar disturbance

in rising from a sitting position on the floor. The patient turns so as to support himself upon his feet and hands; he then places one hand after the other upon his lower limbs, gradually by raising his hands pushing his trunk upward, and in this way climbs up on his lower limbs to the erect position. While not pathognomonic of progressive muscular dystrophy, it is seldom seen in any other affection.

Landouzy and Dejerine believed that the muscles of respiration escape, and in their first case with necropsy the diaphragm was normal, but in a case studied by the writer it was intensely atrophied. Duchenne observed clinically atrophy of the muscles of respiration, and it seems probable, from the study of Bunting, that the heart muscle may be affected.

The belief in the escape of the muscles innervated from the bulb, except those affected in the infantile form, has been shattered by Hoffmann. This investigator has shown that the disease may begin as myopathic bulbar paralysis, and remain as such for some time, and that external ophthalmoplegia may be a part of the symptom complex. Marie observed one case of progressive muscular dystrophy in which the orbicularis palpebrarum muscles were feeble, but the lids could be closed. Double ptosis was pronounced and the muscles of mastication were very weak. It is questionable if any group of muscles possesses more than a relative immunity.

Mental symptoms are not pronounced, but the tendency to degenerative change in the muscles seems to be associated with a form of abiotrophy of the brain; the child, while not imbecile, usually lacks the capacity for high mental development.

Atrophy of bone has been observed in a few cases (Lloyd, Marie, Cruzon, Spiller, Schlippe). In a case reported by the writer the bones of the face as well as those of the limbs were atrophied. An x-ray examination showed that the humerus and scapula on the left side were smaller than the corresponding bones on the right side. The left acromion process and glenoid cavity were unusually small, and the head of the humerus appeared as though it were dislocated forward. The humerus seemed to be deficient in lime salts. The ribs on the left side were larger than those on the right side.

Scleroderma occurs with muscular dystrophy, and is regarded by Meigs as not infrequently associated with it.

Various types of progressive muscular dystrophy have been recognized: (1) The pseudo-hypertrophic; (2) the infantile (Duchenne); (3) the scapular, or juvenile (Erb); (4) the facio-scapulo-humeral (Landouzy-Dejerine); (5) the hereditary (Leyden-Möbius).

The *pseudo-hypertrophic* is the form best known, as it was the earliest recognized (Duchenne), although the juvenile is of frequent occurrence. The implication of the face is extremely uncommon in America, and, indeed is seldom seen in other countries than France. This form affects especially the muscles of the calves, and is more common in males. One or both calves may be affected, and where only one is involved the implication of the other is usually only a question of time. Later, the enlargement may disappear when the atrophy has become intense.

The *facio-scapulo-humeral* type includes the infantile type of Duchenne

and the juvenile type of Erb. The muscles of the shoulder girdle are wasted, also those of the upper part of the upper limbs (deltoid, biceps, triceps, brachialis anticus), as well as the muscles of the neck and face. The eyelids cannot be closed, so that the appearance of bilateral facial paralysis is presented on superficial examination, the lips cannot be puckered as in whistling, and they are usually prominent because of pseudo-hypertrophy, the corners of the mouth are drawn outward in laughing instead of upward, and the muscles of the cheeks and those of mastication may be much wasted. The peculiar appearance of the face has received the name of *facies myopathique*.

Infantile form is an unfortunate designation, as the atrophy may commence in adult life, or even at an advanced age. It is often associated with atrophy of the rest of the trunk and of the lower limbs, and sometimes may follow the latter. This form has interested the French especially. Duchenne was the first to clearly separate this type clinically; he called it progressive muscular atrophy of childhood, and believed that the commencement in the facial muscles never occurred in the adult. He regarded it as the same disease as the progressive spinal muscular atrophy of adults, and called it "*atrophie musculaire progressive de l'enfance*."

The *hereditary* type (Leyden-Möbius) hardly deserves recognition as distinct from other forms. It begins in the lower limbs, as does the pseudo-hypertrophic, ascends from the leg and thigh muscles to those of the pelvic girdle and trunk, but is not associated with pseudo-hypertrophy. It shows very pronouncedly the hereditary tendency. It is dystrophy on an hereditary basis, as are all the other forms. Only a few distinct cases of this type are on record, and Leyden himself reported only one; this, according to Erb, was merely the juvenile form, with predominance of the involvement of the lower limbs. The pseudo-hypertrophic form may, by disappearance of the fatty tissue, change into the Leyden-Möbius type. The latter has, therefore, merely historical interest, and can no longer be recognized.

The type of Zimmerlin consists of atrophy of the proximal parts of the limbs and upper part of the trunk but may begin in the lower limbs and be associated with atrophy of the face. It begins usually about puberty or a little later, but the type is not sharply defined and is closely allied to Erb's juvenile form.

**Diagnosis.**—The clinical appearance usually is so well defined that the diagnosis is not difficult. Babinski observed hypertrophy of a limb developing after typhoid fever, and this condition may resemble muscular dystrophy. Hypertrophy may occur also in syringomyelia. Lipomatosis may be found in the limbs paralyzed in acute anterior poliomyelitis, and may then present the appearance of pseudo-hypertrophic muscular paralysis. Multiple neuritis may be confined to the thighs, as observed by Patrick. The subjective and objective sensory disturbances usually make the diagnosis easy, but in a later stage where the symptoms are chiefly or entirely motor, difficulty in diagnosis might arise.

The resemblance to the Charcot-Marie-Tooth type has already been mentioned, and so soon as this type is enlarged to include cases in which

the atrophy is not confined to the distal portions of the limbs, or atypical signs such as implication of facial muscles occur, the diagnosis becomes questionable.

*Anterior poliomyelitis* may resemble in its late stages progressive muscular dystrophy, especially if the abdominal muscles be paralyzed, but when a history of the onset can be obtained the diagnosis is easy.

In rare instances *myasthenia gravis* may have some resemblance to progressive muscular dystrophy, but the diagnosis should be easy.

The greatest difficulty may occur in distinguishing between the myelopathic and myopathic forms, and in certain cases the decision must be left in doubt, at least for a time.

Disease of the spinal column or of the lumbar muscles may cause the patient to rise from the sitting posture on the floor much as does one afflicted with progressive muscular dystrophy. Congenital defect of muscle may cause difficulty but the defect is not progressive.

**Prognosis.**—Muscular dystrophy is very chronic in its development, and not likely to cause death unless vital muscles become implicated. The afflicted person may have diminished resistance to other diseases.

**Treatment.**—Overexertion is to be avoided. A moderate degree of massage and electricity may be useful in the early stage when only a few muscles are wasted. Exercise should be recommended, as it is probable that muscle will not waste so rapidly if it be employed in such a manner that use does not become abuse. It seems doubtful whether we have any means of influencing the progress, but in certain cases massage, passive movements, and electricity seem to be of decided benefit.

### CHAPTER III.

## THE COMBINED SYSTEM DISEASES OF THE SPINAL CORD.

By COLIN K. RUSSEL, M.D.

COMBINED disease of both afferent and efferent systems of fibres in the spinal cord is found in many conditions, although not all of these can be regarded as true system diseases. In meningomyelitis, myelitis, and multiple sclerosis, the involvement of several of the spinal tracts may, with the subsequent ascending and descending degeneration, give the clinical and anatomical picture of a combined sclerosis. Such a condition has been described also in cases of arteriosclerosis and following injury with contusion of the spinal cord. In these cases we have rather a pseudo-systemic disease of the cord. Clinically, the fundamental character of this form is the spastic weakness of the lower extremities, associated in many cases with ataxia, some loss of sense of position in the extremities or other form of sensory loss. These conditions will be taken up more fully under their respective chapters. It will suffice to state here that ataxic paraplegia of Gowers and spastic paraplegia of Strümpell should be regarded as symptoms only, and not as definite clinical entities. In the majority of cases showing these symptoms in young or middle-aged individuals, with the exception of the subacute conditions to be taken up later, they are merely the early evidence of disseminated sclerosis.

In *tabes dorsalis*, while the disease is usually confined to the posterior columns, occasionally the direct cerebellar tract also shows degenerative changes following on the destruction of the cells in Clark's column (Kattwinkel, Oppenheim) without causing recognizable symptoms. Not infrequently also, as was first observed by Erb in advanced cases of *tabes*, degeneration of the upper motor neurone may occur, giving rise to associated sclerosis of the crossed and direct pyramidal tracts—a true systemic combined sclerosis in some cases—while in others, and according to Crouzon<sup>1</sup> more frequently, it is a pseudo-systemic sclerosis, due to involvement of the lateral columns by disease spreading in from the periphery of the cord from lesions of the lymphatic and circulatory systems. Clinically we have then the symptoms of a spastic paraplegia superimposed on the ordinary tabetic features. If, on the contrary, these myelitic lesions involve the lumbar enlargement and destroy the root zone of the posterior columns, we will have a flaccid paraplegia, and the only direct evidence of involvement of the pyramidal tract may be the presence of Babinski's extensor plantar response.

In general paresis combined degeneration of the posterior lateral columns was first observed by Westphal, and has since been confirmed

<sup>1</sup> *Paris Thesis*, 1904.

by many other observers. In the combined statistics of Tuczek, Furstner, Sibelius, and Crouzon, comprising 200 cases, combined sclerosis was present in 96 cases, that is, in 48 per cent.

### SUBACUTE COMBINED SCLEROSIS OF THE SPINAL CORD.

**Etiology.**—Subacute combined sclerosis occurs usually in individuals in the fourth or fifth decade, although some cases have been reported as occurring in the third and a few in the sixth. Males are just as frequently affected as females, and there is some neuropathic hereditary tendency present in many cases. The writer has recently had under his observation two cases of this condition associated with severe anemia in patients whose mothers were sisters and whose fathers were brothers. Other similar cases of the disease occurring in families have been reported. More important and more constant than this is the almost invariable evidence of some toxic condition, some wasting disease, prolonged suppurative process, chronic digestive disturbances with diarrhoea, or acute or chronic infection. Syphilis apparently plays no important part as an etiological factor. The blood changes so commonly found in these cases are more often of the nature of a secondary anemia than of the pernicious variety, and are probably associated conditions rather than etiological factors, certainly in the majority of cases.

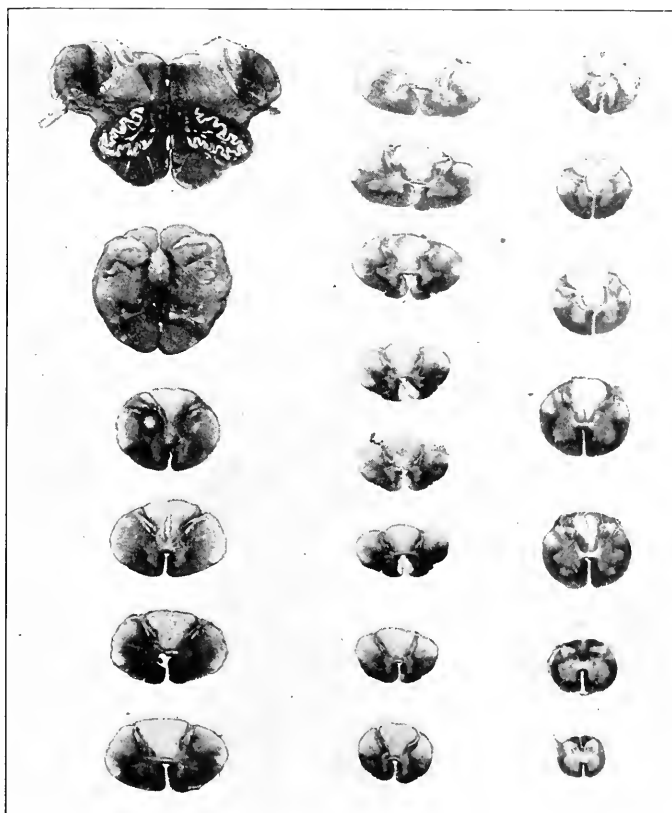
**Pathology.**—The cerebrum, cerebellum, and pons in the great majority of cases show no lesions. Externally the spinal cord and membranes are normal except for an anemic appearance in some cases. Macroscopically, on section the grayish discoloration of sclerosis may be evident in the posterior columns, and sometimes in the region of the crossed pyramidal tract. In many cases this becomes evident only after hardening in Müller's fluid. Microscopically, with the Pal-Weigert method, the most extensive lesions are found in the mid-dorsal region of the cord. At this level almost the whole of the white matter, especially the peripheral parts, may be involved, leaving only the gray matter and a small area of white matter immediately surrounding it perfectly normal. Endogenous and exogenous fibres are alike affected.

The posterior columns, which are usually most affected, may be completely sclerosed except for a small area immediately on the inner border of the gray matter of the posterior horns. At higher levels of the cord the lesions diminish progressively and tend to be limited to the posterior columns, the direct cerebellar tracts, and the pyramidal tracts, both the direct and crossed. Especially in the upper dorsal and lower cervical segments is the degeneration in the region of the direct pyramidal tracts marked. The degeneration in the pyramidal tracts gradually diminishes, and usually has disappeared at the medullary level, although occasionally it can be followed up into the pons by the Marchi method. The degeneration of the direct cerebellar tract is followed by the same method into the inferior cerebellar peduncle, and that of the upper portion of the ventral cerebellar tract (Gowers) has been traced into the velum medullare anterius.

Below the mid-dorsal region the diffuse destructive lesion diminishes



PLATE III



Subacute Combined Sclerosis.



progressively until in the lumbar region the degeneration is confined to the crossed pyramidal and posterior columns. Marchi's method reveals lesions similar to those already described, but somewhat more extensive. Lissauer's tract is not affected, even in the mid-dorsal region, where the periphery of the cord shows marked destructive changes. The cells of the gray matter may show degenerative changes (Putnam, Dana), but, as a rule, these are not at all marked. The nerve roots, especially the anterior ones, were more or less affected in Putnam's cases, but in only one of the cases, reported by Russell, Batten, and Collier,<sup>1</sup> could degenerative changes be demonstrated in the peripheral nerves.

The vessels in the cord, in the affected areas at least, may be engorged and their walls in some cases thickened, but no evidence of thrombosis has been observed. In other parts of the cord they may be quite normal. Minute hemorrhages may be present, associated with the anemia, but having no etiological relationship to the degenerative changes. Russell and his fellow observers are of the opinion that there are two distinct processes at work, first, a system degeneration, and secondly, a focal destructive lesion. With regard to the latter, the first step is a swelling of the medullated sheaths, going on to fatty degeneration, with disappearance of the axis cylinders, absorption of the degenerative products, and later, overgrowth of fibrous tissue. There was in Dana's cases no evidence of any true inflammatory process. As to the former—the system degeneration—the long tracts of the cord exhibit well-marked degeneration and sclerosis. Generally there is clinical evidence to show that the combined sclerosis exists for some time before the onset of the diffuse lesion. This latter always affects the lower cervical or dorsal part, and is usually a warning of an early exitus.

**Symptoms.**—While Risien Russell has divided the course of the disease into three stages, further observations show that these are not always so evident. For the purpose of description, however, they will be retained.

1. The period of mild spastic or ataxic paraplegia. The onset of the disease is in most cases slow and insidious, although in some instances quite acute, being ushered in by headache, vomiting, pyrexia, and malaise. In others it is the symptoms of anemia, palpitation, fainting attacks, and so on, that bring the patient under observation. The first symptoms complained of, attributable to the involvement of the nervous system, are subjective feelings of numbness and tingling in the lower extremities with perhaps some stiffness. These are never severe at first, but soon signs of slight spasticity and ataxia develop. The lower extremities are clumsy, feel stiff and cold, and the toes stick to the floor a little in walking. At this period there is no loss of control of the sphincters and no evidence of involvement of any other part of the nervous system. One finds on examination some impairment of the sense of position in the lower extremities when they are moved passively, with increased reflexes, and the extensor plantar response (Babinski). As a rule, later, though sometimes coincidentally, the same symptoms appear in the upper extremities. This period of ataxic paraplegia occupies half or three-quarters of the whole duration of the disease.

<sup>1</sup> *Brain*, 1900, xxiii, 39.

2. This period is one of complete spastic paraplegia, with considerable loss of sensibility in all its forms in the lower extremities and lower part of the trunk. The transition from the first to the second stage is, as a rule, abrupt; often in the course of twenty-four hours the patient loses the power of standing or walking, owing to the absolute loss of sense of position in the limbs, and not from any loss of motor power, which is usually not much impaired. Throughout this second stage the motor power diminishes gradually, as a rule, but is not entirely lost until the third stage is reached. The upper extremities are affected also with the loss of sense of position, and often show tabetic athetosis. The loss of cutaneous sensibility is at first peripheral in type, but as it extends upward becomes segmental in distribution, and its upper limit on the trunk is usually well defined, it may extend as high as the area of distribution of the sixth cervical segment. Girdle sensation and lightning pains are not uncommon, and frequently the patient complains of a severe constant dragging pain beneath the lower costal margin. Herpes may occur, and Russell reports one case with cutaneous hemorrhages similar to those observed by Strauss in tabes. The mental condition remains unimpaired; and only occasionally is there any implication of the cranial nerves. There may be some loss of sphincter control; trophic disturbances are not present. The deep reflexes are all exaggerated, and there is extensor plantar response (Babinski).

In some, though by no means in all cases, the symptoms of involvement of the nervous system are associated with the evidences of anemia. There is constantly some irregular pyrexia. The average duration of this stage of the disease is about five weeks, then in the course of a few days the third period follows.

3. The third stage, that of *flaccid paraplegia*, is characterized by an absolute flaccid motor paralysis of the lower extremities. The trunk muscles also, with the onset of the third stage, show complete paralysis below a segmental level, and Beever's sign (*i. e.*, the riding up of the umbilicus on the attempt to raise the head from the bed, due to the paralysis of the lower segments of the recti abdominis) can, as a rule, be observed in this stage. Subsequently the upper abdominal and spinal muscles and the lower intercostals are progressively attacked. There is absolute loss of sensibility, loss of the tendon-jerks, rapid muscular atrophy, and rapid loss of excitability of the muscles to faradic stimulation; oedema of the lower extremities and the lower part of the trunk with loss of sphincter control follows.

With the onset of these signs there is an elevation of temperature, with general malaise, drowsiness, anorexia, and general asthenia. If anemia be already present, it is much increased. In the lower limbs and trunk, then, the wasting is general, but in the upper extremities it is of a segmental character, the intrinsic muscles of the hand being first affected, then the ulnar flexors, radial flexors, extensors of the wrist, extensors of the elbow, supinators, and fifth cervical group in that order. In some cases the diaphragm may become paralyzed. Involvement of the cranial nerves is very exceptional. Mental disturbance is usually seen in this stage, and there may be convulsions. Trophic changes and bedsores are apt to occur in spite of the most careful treatment. The deep reflexes are lost with the

onset of flaccidity, while the extensor plantar response (Babinski) remains present. The duration of this period is about six weeks, though exceptionally it may last several months, death being due either to sudden syncope or respiratory failure.

One of the characteristic features of this disease is its progressiveness; neither remissions nor amelioration of the nervous symptoms ever occur, even though the general condition may improve somewhat. There is always some irregular fever not to be accounted for by associated conditions, such as cystitis, bedsores, etc. The duration of the disease varies from a few months to two or three years. The relation of the anemia to the cord lesion is not invariable; in some cases it precedes the appearances of the nervous symptoms, in others it may appear coincidently with the nervous symptoms and increase progressively with them; or it may be present only at the end of a prolonged third stage when emaciation becomes extreme, or there may be no anemia during the whole course.

The essential nature of this process is evidently a primary nerve degeneration. The same toxin that brings this about may cause more or less severe disturbance of the blood-forming elements. Its action on the nervous system affects first, as one would expect, those long tracts whose mere length must necessitate a more elaborate nutrition for their maintenance. The circulatory conditions probably affect the preterminal diffuse lesion in the dorsal region in the same manner that acute transverse myelitis usually affects the dorsal region of the cord.

**Diagnosis.**—In the first stage this disease must be differentiated from the early condition of disseminated sclerosis, that is, the ataxic paralytic stage. Of course, when the classical advanced picture is present with nystagmus, intention tremor, scanning speech, etc., no special art is required. In the early stages, however, the similarity may be striking. Disseminated sclerosis usually comes on at an earlier age than does subacute combined sclerosis. If anemia be present, it should be suggestive; and the subjective and objective sensory disturbances are much more marked and persistent in subacute combined than in disseminated sclerosis. The subacute progressive character of the one disease with its preterminal emaciation is of course quite different from the slow and prolonged course of the other.

In the later stages, with the flaccid paraplegia, the condition may simulate peripheral neuritis, myelitis, tumor of the cord, or tabes. The history of spastic gait, the involvement of the sphincters, the paralysis and loss of sensibility extending to the trunk and its nerve-root distribution, but especially the extensor plantar response, should be sufficient to make a diagnosis from multiple neuritis. Acute myelitis is of course excluded by the prolonged history. The absence of severe root pains and the distribution of the physical signs in unequal degree in the lower and upper limbs should exclude a tumor in the spinal canal.

The condition of flaccidity, anesthesia, loss of deep reflexes, and complete incontinence often may suggest *tabes*. The history of lightning pains in the second stage may make it more suggestive. The previous history of spasticity, changing rapidly to flaccidity, the absence of the Argyll-Robertson pupil, the great loss of motor power, the rapid muscular wast-

ing and the loss of faradic excitability and the extensor plantar response, should prevent error.

Enough has been said in describing the symptoms and course of the disease to make it evident that the *prognosis* is extremely bad. Any *treatment*, to be at all effectual, must be prophylactic. Attention to the general nutrition in all such debilitated and cachectic conditions is, of course, necessary. The patient should be at rest in bed in the early stages, to prevent as far as possible any exhaustion of neurones whose vitality and recuperative power are impaired by the presence of the hypothetical toxic matter in the circulation. Iron and arsenic are indicated to combat the anemia, and the usual dietary and hygienic measures.

### FRIEDREICH'S ATAXIA. MARIE'S HEREDITARY CEREBELLAR ATAXIA.

Friedreich's ataxia is a disease, probably of abiotropic nature, characterized by a slow and progressive incoördination of the four limbs. The disturbance begins in the lower limbs, extends to the trunk and upper limbs, and finally involves the tongue, larynx, and eyes. The disease generally affects several members of the same family. It is named after the famous Heidelberg professor, who first described the disease.<sup>1</sup> He looked upon it as an hereditary form of *tabes dorsalis*. One must remember that Duchenne, of Boulogne, had recently described his progressive locomotor ataxia, so that sclerosis of the posterior columns of the cord and *tabes* were synonymous in the minds of most neuropathologists of that time. Schultz, of Bonn, first drew attention to the associated sclerosis of the lateral columns and posterior horns. In Rutimeyer's publication,<sup>2</sup> in 1883, the view was advanced that we have in this disease a primary systemic combined sclerosis developing on an hereditary foundation.

In 1893 Pierre Marie<sup>3</sup> gave the name of *hereditary cerebellar ataxia* to a syndrome characterized by ataxic gait and cerebellar incoördination. He took this stand on the study of sixteen cases of familial disease, published by Fraser, Nonné, Sanger Brown, Klippel, and Durante. The main differences between this type and the type of Friedreich are, in the first place, a more definite hereditary influence in the former cases, although this is accounted for by the fact that the onset being much later, in fact, about the third decade, allows an opportunity for procreation, which is not possible in the patient suffering from the usual Friedreich type, in which the onset is before or about puberty; and secondly, the occurrence of ocular disturbances, the retention and even exaggeration of the knee- and ankle-jerks, and the absence of those trophic disturbances, the *pes cavus* and curvature of the spine, so characteristic of the Friedreich type. Marie considered that the syndrome bearing his name and Friedreich's disease were possibly different types of one and the same affection, due to an initial heredo-dengeneracy of the nervous system. He was of the opinion that the lesion in the cases of his type was localized more definitely in the *cerebellum* itself.

<sup>1</sup> *Virchows Archiv*, 1863, xxvi, 391, 433; 1864, xxvii, 1; 1876, lxxviii, 145; 1878, lxx.

<sup>2</sup> *Ibid.*, 1883, xci, 106.

<sup>3</sup> *Semaine Médicale*, 1893, xiii, 444.

The appearance of cases which clinically are intermediate between the ordinary Friedreich type and Marie's hereditary cerebellar ataxia, and moreover, the microscopic examination in Sanger Brown's and similar cases, which reveal no particular affection of the cerebellum, but a condition very similar to that found in the ordinary spinal type of the disease, justify us in including Marie's type as a form of the same disease. In fact, no dividing line can be drawn between the two types either clinically or pathologically.

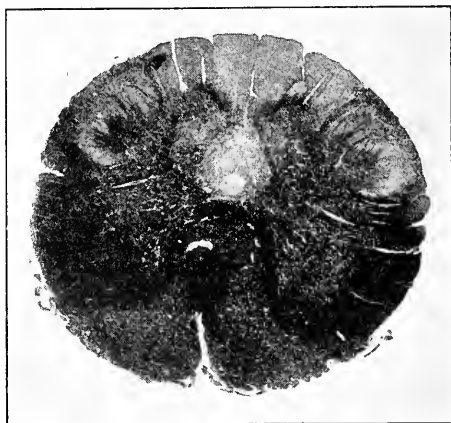
**Etiology.**—The disease is evidently what Sir William Gowers describes as an abiotrophic condition, a congenital inherited and inherent lack of vitality in certain parts of the nervous system. Other stigmata of degeneration are, as a rule, not lacking, and cases have been reported associated with infantilism, feminism, maldevelopment of the testicles, and so on. The congenital tendency is shown by the usual occurrence of the disease in several members of the same family, its onset at approximately the same age, and the similarity in the type of the developed disease in the different members affected. A neuropathic diathesis is present in most cases, as is frequently shown by the occurrence of hysteria, migraine, epilepsy, or insanity in collaterals or ancestors. Alcoholism in the parents was first insisted upon as an etiological factor by Friedreich, his first six cases being members of two families the fathers of which were drunkards and the children conceived in drunkenness. Quinke, Rutimeyer, Althaus, Everett Smith, Destree, and others have related analogous examples. Hereditary lues and tuberculosis may be predisposing causes. Consanguinity in the parents has been reported in many of the cases. Any fall, injury, or infectious disease may appear to determine the onset, and any cause acting as a depressant to the vital forces produce a rapid advancement of the symptoms. The influence of sex is not noticeable, females being as frequently affected as males.

**Pathology.**—In many cases of Friedreich's ataxia the cord has been found to be markedly smaller than normal. This atrophy affects chiefly the posterior columns. Anomalies of structure may be present; thus Friedreich found a supplementary central canal in three cases. The cerebellum has been reported as smaller than normal in some cases. The cerebrum is not affected. There is occasionally some thickening of the membranes, particularly over the posterior aspect of the cord, sometimes, as in Mingazzini's case, more accentuated in the lumbar region.

On section of the cord, microscopically there is found a posterolateral sclerosis affecting the columns of Goll and Burdach, the posterolateral or direct cerebellar tract, and in many cases the anterolateral or Gowers' tract. The crossed pyramidal tract is constantly affected in some degree at least, and in a few cases the direct pyramidal tract has also shown degenerative changes. In the gray matter, cellular lesions in the column of Clarke have been invariably found in association with the degeneration of the direct cerebellar tract. Associated with atrophy of the muscles of the extremities, atrophy of the anterior horn cells and thinning of the reticulum has been described by some observers.

Lissauer's tract shows degeneration, which may be chiefly in the lumbar region. The posterior roots in most of the cases examined recently show a

FIG. 8



Friedreich's ataxia. Lower medulla.

variable amount of degeneration. The spinal ganglia are, as a rule, practically normal, although atrophic degeneration of the nervous elements with connective-tissue hyperplasia has been found by some observers. The peripheral nerves show no constant change. The sclerosis of Goll's column, is, as a rule, complete, and can be followed up to its nucleus in the medulla, while that in Burdach's column is not so complete and varies in extent at different levels. In Mingazzini's case, and in those reported by Du-

FIG. 9

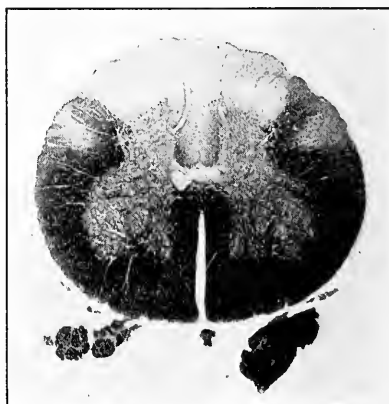


Friedreich's ataxia. Cervical region.

mon, Philippe, and Oberthür, the lesions of the posterior roots seem to correspond more or less closely with the lesions in the posterior columns.

There are always some intact fibres left along the inner border of the posterior horns and immediately posterior to the gray commissure. Dejerine first called attention to the formation of what he termed "*tour-billions*," *i. e.*, small twirls of neuroglial tissue, in sclerosed areas of the posterior columns. These probably have no particular significance, and

FIG. 10



Friedreich's ataxia. Lumbar region.



may be found in other long-standing sclerotic conditions in the cord. The sclerosis of the direct cerebellar tract follows its usual course, beginning in the lower dorsal region and gradually becoming attenuated toward the medulla. Gowers' tract shows an almost constant sclerosis, which according to Marie is invariably present if the disease be sufficiently advanced. The sclerosis of the crossed pyramidal tract is not as dense as in the posterior columns, and may be present only in the lumbar and lower dorsal regions, though in more advanced cases it may be visible even at the upper cervical level.

Marie, in his type, termed hereditary cerebellar ataxia, basing his views on the findings in Fraser's and Nonné's cases, was of the opinion that the underlying pathological lesion here was an atrophic condition of the cerebellum. In Fraser's<sup>1</sup> cases the cerebellum was reduced to less than half its normal weight, its cortex was little more than half the normal thickness, very few Purkinje cells remained in it, and these were atrophied and shrunken. There was no macroscopic degeneration in the spinal cord. In Nonné's<sup>2</sup> case the cerebellum was abnormally small, but no further lesion was demonstrable. Later autopsies, however, performed by Meyer and by Barker on three cases of Sanger Brown's remarkable series, have not altogether confirmed these observations. Adolph Meyer<sup>3</sup> found no circumscribed cerebellar lesion; Barker<sup>4</sup> found no marked diminution in the size of the cerebellum, although it, with the cord, medulla, and pons looked small in proportion to the cerebrum, which itself was small. The microscopic study in both cases revealed a marked degeneration in the gray and white matter of the spinal cord, medulla oblongata, and cerebellum. The degeneration involved chiefly the nerve cells and fibres of the centripetal paths, including the posterior columns, the dorsal nucleus of Clarke, the direct cerebellar tract with its continuation in the restiform body. There was practically no change in the cerebellum beyond a possible diminution in the number of cells in the nucleus dentatus and nucleus fastigii.

**Symptoms.**—Following the order taken up in the preceding section, that type of the disease described by Friedreich will be first discussed, then the so-called hereditary cerebellar ataxia of Marie, and finally, attention will be directed to the numerous intermediate forms between these clinical types.

In the type of Friedreich, the disease commences at an early age, usually between four and ten, or at puberty; in exceptional cases the onset may be later. It is probable that in many cases the first symptoms pass unobserved until after some infectious fever or injury of some kind, when a slowly developing ataxia of the lower extremities is noticed. The child walks somewhat hesitatingly, with legs rather far apart, and attempts to balance himself with the arms; the feet are brought down in rather a stamping fashion, the paces are irregular, with some deviation in the line of march, the picture resembling somewhat a partially intoxicated person. The child is not steady on his feet, but falls more readily than other chil-

<sup>1</sup> *Glasgow Medical Journal*, 1880, vol. i.

<sup>2</sup> *Arch. f. Psych.*, 1891, xxii, 283.

<sup>3</sup> *Brain*, 1897, xx, 276.

<sup>4</sup> *University of Chicago, Decennial Publication*, 1903, vol. x.

dren. In standing alone there is usually some unsteadiness, and this, as a rule, is not increased by closing of the eyes; so that, although Romberg's sign has been reported in some cases, it is not typical of the disease.

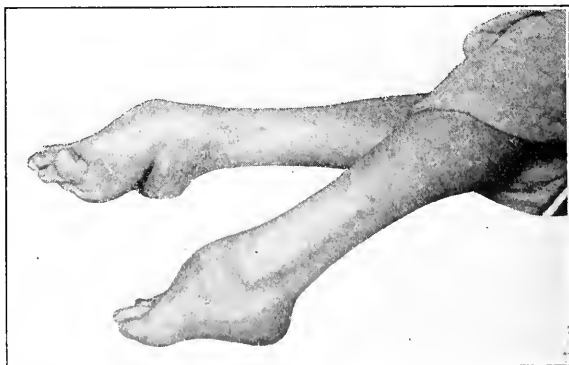
FIG. 11



Friedreich's ataxia, showing the spinal curvature.

On further examination in this early stage the cranial nerves are not found affected; speech may in exceptional cases be somewhat slow and drawling quite early, but, as a rule, the defects in speech and nystagmus are observed later. There is no atrophy or loss of power to be made out

FIG. 12



Friedreich's ataxia, showing the typical deformity of the feet.

in the muscles. Their tone is usually about normal; there is no impairment of the sense of position of the muscles, nor can any loss of any other form of sensibility be demonstrated. The knee-jerks in this early stage

are most commonly lost, but are often retained and may even be active. The extensor plantar response (Babinski) may or may not be present. There is very often, even at this early stage, some scoliosis of the spinal column, and there may be evidence of the beginning of the club foot so typical of the fully developed disease. The bladder functions are not disturbed, nor do they usually suffer.

Gradually the disease progresses, and in the course of a few years the typical picture is developed. The ataxia affects the muscles of the upper extremities, trunk, and neck. At first it may be observed associated with voluntary movements only. If the child has already learned to write, it is noticed that his penmanship, instead of improving becomes awkward and clumsy. Articles are knocked over in the attempt to grasp them. The movements are associated with a swaying, sometimes almost choreiform, jerkiness, until finally the patient may not be able to convey a glass of water to his mouth. The ataxia is of the nature of a motor incoördination, due to the imperfect fusion of simple contractions, which compose voluntary movements. The disturbance of speech and the nystagmus are of a similar nature. A condition of static ataxia also develops, caused by this same defect in the permanent, regular, harmonious innervation of those muscles and their antagonists, which are necessary for the maintenance of equilibrium and the upright position of the body. This may be first noticed in the muscles of the neck, causing a jerky, irregular, almost tremor-like movement of the head. Later, as the disease progresses and the trunk muscles become affected, the patient has difficulty in maintaining the sitting position without support, on account of the irregular movements of the trunk on the pelvis. Associated with the ataxia there may be a certain amount of muscular weakness, but this is not usual except in advanced cases, when the patient becomes confined to a chair or bed-ridden. In this stage and sometimes even earlier, atrophy of the muscles of the extremities may be present, associated with contractures and malposition. Faradic excitability of the muscles may be lost and the reaction to galvanism diminished. The writer has observed this also in the muscles of the lower extremities where there was no apparent atrophy.

The cerebral functions are commonly not impaired, although the general facial expression may suggest extreme stupidity; this is more often due to simple apathy and indolence. Impulsive laughter has been described in some cases. Excessive salivation has been known to occur. Speech becomes slower, hesitating, and jerky; articulation is imperfect, some syllables are prolonged, others forced out quickly in an explosive manner, as if the ataxia had affected the muscles of articulation. In a typical case the optic nerves are not affected, nor is the Argyll-Robertson pupil a symptom. Nystagmus, associated with lateral movements of the eyes, is not infrequent. Disturbances of respiration may be caused by the involvement of the abdominal muscles in the ataxia, and if the medullary centres be affected a definite dyspnoea results.

The sensory system is, as a rule, unaffected. Lancinating pains were present in some few cases reported, but are not common. Cramp-like pains in the limbs are perhaps rather more frequently complained of. Only in exceptional cases is there any impairment of sensibility, and then

usually late on in the disease and of an indefinite incomplete type, being confined to the more distal parts of the extremities. There is rarely any loss of the sense of position of the muscles, although it has been observed in exceptional cases (Oppenheim). Visceral crises never occur. The tendon-jerks gradually diminish, disappearing first in the lower extremities. The presence of the extensor plantar response (Babinski), with the absence of the knee- and ankle-jerks, is typical.

Vasomotor paresis occurs, so that in many cases the feet and ankles are congested, reddish blue in color, and clammy. Lateral curvature of the spine is almost always present, commencing early in the disease and often reaching an extreme grade. A peculiar deformity of the feet is commonly seen, namely, a condition of pes equinovarus, with hyperextension of the proximal phalanx and flexion of the distal phalanx of the great toe. In families affected with the disease this sign has in some cases been what they themselves looked for, knowing from hard experience what the ultimate outcome would be when it once made its appearance.

In *hereditary cerebellar ataxia* the same familial character is present, but, as has been remarked above, the hereditary tendency is more evident on account of the disease commencing only in the third or fourth decade, and thus permitting the victims to marry and produce offspring. In the celebrated series reported by Sanger Brown,<sup>1</sup> twenty-three members of one family, spread over four generations, were affected. Females are perhaps slightly more frequently affected than males; and Sanger Brown remarks that the disease was inherited more frequently through the girls than through the sons of the family. Occasionally the disease may skip one or even two generations, appearing perhaps in several members of the third.

The disease commences with a slowly progressive ataxia, more or less marked in walking and standing; lancinating pains in the legs may be present. After some time the ataxia affects the upper extremities, and soon disturbances of speech are noticed, and the patient complains of difficulty of vision. Tendon-jerks are increased, instead of being diminished or lost, as we find in the Friedreich type, and occasionally other evidences of spasticity in the lower extremities are seen. The extensor plantar response is present. Only in a few cases has there been any disturbance of cutaneous sensibility. Frequently mental weakness develops in the course of the disease. Disturbance of deglutition and impairment of the control of the sphincters occur only very rarely. The disease is essentially progressive, but may present remissions. Some intercurrent disease, probably years after the onset, is the usual cause of death.

The disturbance of function of the lower extremities is absolutely analogous to what is observed in Friedreich's disease. The gait and station are similar to that already described, starting gradually, progressing steadily, increasing with fatigue. Frequently in the early stages the unsteady staggering gait leads to the accusation of inebriety. Closing of the eyes does not increase the unsteadiness in standing to any extent. The upper extremities are affected to a much less degree and later in the course of the disease. As in Friedreich's type, it is the more delicate movements that

<sup>1</sup> *Brain*, 1892, xv, 250.

are most interfered with in the early stages; later even the gross movements may become equally affected. Static ataxia also develops; there is no muscular weakness.

The signs of spasticity of the lower extremities, on which the differentiation of this type from Friedreich's is based, to a great extent consist in increased knee-jerks and in some cases ankle clonus, contrasting with the absence of knee-jerks in the typical picture of Friedreich. The extensor plantar response (Babinski) is present. The tone of the muscles is increased, in some cases to such a degree that it can be overcome only with difficulty.

Apart from the lancinating pains occasionally present in the earlier stages, there is seldom any disturbance of sensibility. The sense of position in the muscles was disturbed in Klippel and Durante's cases, but, as a general rule, it is unaffected.

In the domain of the cranial nerves ptosis is sometimes present, nystagmus is a common symptom, and frequently one finds paralysis of one or other of the external ocular muscles. The pupils often show a diminished or even an abolished reaction to light and occasionally disturbance of the reaction to accommodation. The visual field often shows some diminution in size, the color vision may be impaired. Primary optic atrophy is not uncommon. The disturbance of speech is similar to that described in Friedreich's type. A peculiar overaction of the muscles of the face, especially marked on the attempt to speak, is frequently observed. Spinal curvature and club foot are not, as a rule, included among the symptoms of cerebellar ataxia; they do sometimes occur (Londe, Muira, Rossolimo).

Numerous forms intermediate between these two types have been observed. Thus, for instance, Bonnus reported a case, otherwise conforming to the classical description of Friedreich's disease, in which the onset of symptoms began after the twenty-fifth year. In a brother and sister, reported by Baumlin, typical cases of the cerebellar ataxia type, with the ataxia, pains, convergent strabismus, optic atrophy, and increase of the patellar and other tendon-jerks, the onset in the boy during childhood, in a girl at the age of fifteen years. Similar cases to these have been reported by Rossolimo and Lennmalin.

In typical cases of Friedreich's disease the knee-jerks are abolished. Many cases have been observed, however, in which in the early stages they have been preserved, and disappeared only later in the course presumably when the degeneration in the posterior fibres affected the part of the cord interested.

The two types cannot be differentiated clinically by the presence or absence of signs of spasticity in the lower extremities. Erb has reported two sisters whom he regards as examples of Friedreich's disease in spite of normal knee-jerks. Hodge has described three cases of the same type, all presenting marked increase in the knee-jerks. Allen Starr found in one family two of the affected members with loss of knee-jerks, while in the youngest, with quite similar symptoms, the knee-jerks were rather increased. Brock and Tressider report similar instances in which the knee-jerks were present in some and absent in others of the affected members of a family, and it is by no means infrequent. Paralysis of the external

muscles of the eye in otherwise typical cases of Friedreich's disease has been observed by Gowers, Mendel, Omerod, Jaffroy, Anderson, Starr, and others. Optic atrophy was present in the case of two brothers reported by Cohn.

It is evident that no fast dividing line can be drawn between the various forms of this disease. We have here an abiotrophic condition in which the localization of the degenerative process determines the clinical signs.

**Diagnosis.**—In the early stages it is possible that this disease may be mistaken for chorea, especially if there happens to be only one member of the family affected. The family history, curvature of the spine, absent knee-jerks, with the extensor plantar response, should leave no doubt, and of course the progress of the disease renders such an error impossible for any length of time.

*Tabes dorsalis* may be suggested by the ataxia and the loss of knee-jerks, but as a matter of fact there is little danger of such a mistake. The ataxia is quite different. In tabes it is due to a loss of the sense of position of the muscles; in Friedreich's it is entirely a motor incoördination, not increased to any extent in the dark or when the eyes are closed. The gait is quite distinctive; there is not the same overaction of all the movements in Friedreich's ataxia. The difficulty is more in maintaining the equilibrium than in governing the action of the limbs, and the patient does not watch his feet so intently. The age of onset of the disease, the history of antecedent lues, the disturbance of the bladder functions, the lightning pains, girdle sensations, and paresthesias in tabes, with objective sensory disturbances and the Argyll-Robertson pupil, leave no doubt about the diagnosis. Scoliosis and the characteristic club foot form no part of the picture of tabes. The mental condition and absence of Argyll-Robertson pupil exclude juvenile general paresis.

The differential diagnosis between *disseminated sclerosis* and those sporadic cases of Friedreich's ataxia is sometimes difficult, especially when the knee-jerks are not lost in the very early stages. The incidence of the disease in several members of a family gives the clue immediately, and spinal curvature, club foot, and cerebellar gait settle the question. Especially in sporadic cases of the cerebellar ataxic type will the differential diagnosis be difficult. The disturbance of volitional movement, the nystagmus, the speech defect, the increased knee-jerk, and extensor plantar response will suggest disseminated sclerosis most strongly. The difficulty will be much increased if the patient be already in a bedridden condition when he first comes under observation. The muscular incoördination of the upper extremities with no sensory disturbance is similar in the two diseases; increased reflexes and extensor plantar response are common to both. In each there is disturbance of speech, nystagmus, ocular paralyses, and not infrequently optic atrophy. In the cerebellar ataxia, scoliosis and club foot are only exceptionally seen, and never in disseminated sclerosis. The gait, however, is distinctive. In sclerosis it is spastic, progression is direct enough, the paces regular but short, the legs stiff, and the toes, clinging as it were to the floor, are apt to catch on any little obstacle or unevenness.

The steady progression in hereditary ataxia is quite different from the

intervals of remissions even with some improvement followed by exacerbations, which one sees in the disseminated form. In the latter, disturbances of bladder functions are more common. Absence or slowness of the reaction of the pupil to light is more common in hereditary ataxia.

The appearance of the movements, and the family character of the disease in Huntington's *hereditary chorea*, may also give rise to some uncertainty. The presence of nystagmus, ocular palsies, optic atrophy, and especially the state of the reflexes, should remove all doubt. In hereditary chorea mental aberration is the rule sooner or later.

**Prognosis.**—In either type the prognosis is bad. Its steadily progressive character has been sufficiently emphasized. Sooner or later the patient becomes bedridden, but he may live for twenty or thirty years, eventually being carried off by some intercurrent disease.

**Treatment.**—Drugs have no effect on the disease process. Reëducative exercises, as in tabes, may be of some benefit. More might be looked for from the prophylactic treatment of the younger and unaffected members of the family; proper hygienic measures and every care should be taken to improve and maintain their general health.

### COMBINED SCLEROSIS OF THE CORD CAUSED BY VEGETABLE POISONS.

**Lathyrism.**—Lathyrism, according to Sheube, is a disease with a spastic spinal paralytic course, of the nature of an intoxication which is attributable to poisoning by various kinds of grass pea or Papilionaceæ lathyrus.

**Etiology.**—The disease is endemic in certain parts of France, British India, and Algiers. Only the poorer farm laborers are affected, as a rule, and the incidence is much increased in famine years, the poorer classes being forced to live on the cheapest sort of food, such as *Lathyrus sativus*, *L. cicera*, or *L. clymenum*. Men are more frequently affected than women.

**Symptoms.**—The disease commences with general weakness and trembling of the legs, and, according to Brunelli,<sup>1</sup> if the food be not changed, a condition of spasmodic tabes develops in the course of a month or two; the onset may in some cases be even more abrupt. It is characterized by spasticity of the lower extremities, bladder disturbance, paresthesia, and blunting of cutaneous sensibility. There is no muscular atrophy, and no involvement of the cranial nerves. The patient walks with the aid of a long two-handed staff in a typically spastic manner.

Pigs, dogs, and rabbits are also occasionally affected, the disease showing itself by paralysis of the posterior extremities. Birds appear to be immune.

The underlying pathological lesion is a posterolateral sclerosis, but the pathology is still obscure.

**Pellagra.**—It is only necessary to mention here the combined sclerosis of the posterior columns and the crossed pyramidal tracts that one finds in advanced cases of Pellagra and the reader is referred to the proper chapter for the further consideration of this disease.<sup>2</sup>

<sup>1</sup> *Trans. Seventh International Congress, London, vol. ii.*

<sup>2</sup> Volume II, p. 472.

## CHAPTER IV.

### SCLEROSES OF THE BRAIN.

By EDWIN BRAMWELL, M.B., F.R.C.P. (LOND. AND EDIN.).

**Introduction.**—An increase of the neuroglia and mesodermic connective tissue of the central nervous system occurs in a variety of pathological conditions. Proliferation of the neuroglia is met with, for example, in the systemic disorders, whether these be of abiotrophic, ischemic, or toxic origin. A similar neuroglial increase accompanies the systemic degenerations consequent upon interruption of the nerve fibres in some part of their course. A local proliferation of the interstitial tissue may be observed in the neighborhood of inflammatory lesions and old foci of hemorrhage, while a cortical sclerosis is found postmortem in some cases of infantile hemiplegia as a sequel, in the opinion of Strümpell, to a primary acute encephalitis. The neuroglial proliferation in the instances mentioned is either of compensatory origin, the tissue multiplying in order to occupy the place left vacant by the disappearance of the nerve elements, or secondary, in the sense that the change constitutes a later stage in the course of an inflammatory process.

A group of affections remains for consideration—the scleroses proper of the nervous system—in which the neuroglial proliferation, if not a definite primary process, is at least the dominant feature. Disseminated sclerosis, one of the commonest diseases of the nervous system, is by far the most important of these. Diffuse sclerosis, a very rare affection, belongs naturally to this group. Pseudo-sclerosis, another disease of great rarity, may also be included, since it closely resembles clinically the two last mentioned affections, while pathologically it is regarded by some authorities as an early stage of diffuse sclerosis. Miliary and tuberosc scleroses are pathological entities of little clinical interest which call for brief notice.

#### DISSEMINATED SCLEROSIS.

**Synonyms.**—Multiple sclerosis; insular sclerosis; sclérose en plaques.

**Frequency.**—Disseminated sclerosis is one of the commonest organic affections of the nervous system. Its frequency appears to vary in different countries. In Scotland it would appear to be commoner than in the United States (Byrom Bramwell, Jelliffe). E. W. Taylor (1906), who has collected 13 cases with autopsies from the American literature, is of opinion that even in the States it is a common disease, though far less frequent than tabes, and is probably often overlooked. Among 1000 consecutive cases of nervous disease seen by Byrom Bramwell in his private practice, there were 32 cases of disseminated sclerosis and 34 of tabes. Strümpell observes that in a mixed population, to judge from his Erlangen



and Breslau material, disseminated sclerosis is distinctly commoner than tabes. E. Müller, who analyzed 80 cases of disseminated sclerosis from Strümpell's Erlangen clinic, found that during the same period syringomyelia had been diagnosed on 29 occasions.

**Etiology.**—The causes are most obscure, and no factor common to a large proportion of cases has hitherto been demonstrated, although in individual instances indications of an etiological relationship either predisposing, exciting, or augmentative are discernible.

**Sex.**—There is a surprising difference of opinion as to the incidence of sex, in part, at least, to be explained by the hospital accommodation at the disposal of various observers. Thus, among 34 cases observed by Charcot there were only 9 males. In Byrom Bramwell's series of 110 cases, 67 were females. Bruns and Berlin record a preponderance in the female sex. On the other hand, Uhtoff, among 100 cases, met with 67 males, while in Frankl-Hochwart's series of 206 cases there were 140 males. Tredgold, in 1904, collected 373 recent cases from the literature; 200 of these were males as against 173 females. Thus it would appear that the sexes are almost equally liable.

**Age.**—The first symptoms usually develop during early adult life. E. Müller found that in 44 per cent. of his cases the earliest signs appeared during the third decade. Byrom Bramwell met with 55 per cent. commencing during this period, while in 85 per cent. of his cases the disease first manifested itself between the fifteenth and thirty-fifth years. Oppenheim has seen 3 cases confirmed by autopsy in which symptoms were first noticed between the ages of thirteen and fifteen. It is very unusual for the disease to begin after forty years of age.

Instances have been reported in which some of the symptoms appeared to date back to infancy. Schupfer,<sup>1</sup> who in 1902 collected 59 cases reported as commencing in childhood, considers that the diagnosis was often unwarranted. Müller, after discussing the data, came to the conclusion that a true multiple sclerosis in childhood identical with that occurring in the adult has yet to be demonstrated.

**Infections.**—Kahler and Pick indicated in 1879 that insular sclerosis often develops after an acute infective disease. Pierre Marie in his *Diseases of the Spinal Cord* (1892) strongly advocates this view. Of 25 cases which appeared to follow upon an infective malady, typhoid fever was in 11 instances the preceding disease. The apparent onset of the first symptoms after influenza, measles, scarlet fever, diphtheria, smallpox, cholera, etc., has been observed. To account for this, Marie supposes that its cause is "the result of one of those combined infections which are so frequent during the course of the different infective diseases." It cannot be denied that an infective malady sometimes appears to act as an augmentative cause, yet it is extremely doubtful whether it alone is ever the essential factor. The writer's experience has convinced him that in a considerable proportion of these cases, careful inquiry demonstrates that undoubted symptoms of the affection have been previously in existence. The prevalence of influenza during the last two decades has been regarded

<sup>1</sup> *Monatschr. f. Psych. u. Neurol.*, Band xii, Heft 1 und 2.

as possibly accounting for the increase in the number of cases of disseminated sclerosis met with during the same period. Advance in diagnosis offers an alternative explanation. Finally, although it is improbable that the infective diseases actually determine the incidence of disseminated sclerosis, for, in the great majority of cases, there is no history of a preceding illness, it must be admitted that a very pronounced increase in the severity of the symptoms is a common sequel to these disorders.

**Syphilis** plays no causative rôle in relation to disseminated sclerosis. Proof of this assertion is found in the rarity with which a history of syphilis or evidence of that disease is forthcoming (there was a history of syphilis in only 3 of the 206 cases analyzed by Fränkl-Hochwart), the fact that the sexes suffer almost equally, and the circumstance that among a population in which both syphilis and the parenchymatous syphilitic affections are common there is no corresponding increase in the cases of disseminated sclerosis. The writer's experience in this connection is of some interest. In the Out-patient Department of Leith Hospital, which draws its material from a seaport town of 80,000 inhabitants, during a period of four years he met with 62 cases of tabes as compared with 2 cases of disseminated sclerosis.

**Trauma.**—In 1901 Gumbrecht<sup>1</sup> collected 54 cases in which it was claimed that trauma was of etiological importance. Grossmann<sup>2</sup> has reviewed the literature. Von Leyden, Hoffmann, and other observers recorded cases in which they regarded injury as a factor of importance. E. Mendel held, and with this opinion the writer is inclined to agree, that exceptionally injury may act as an exciting cause in an individual who is predisposed.

**Metallic Poisons.**—Oppenheim published observations which suggest that poisoning by lead and other metals may play a rôle. Among 28 cases in which the diagnosis was certain, and in which special inquiry was made into this point, in 11 a history of exposure to possible metallic poisoning by lead, copper, zinc, and other metals was obtained. The rarity of such a possible etiological relationship is generally admitted.

**Hereditary Predisposition.**—Eichorst and Lenot have recorded cases of disseminated sclerosis occurring at an early age, in each instance the mother of the patient having suffered from that disease. In Eichorst's case, which was examined postmortem, the pathological findings are not altogether conclusive. In very rare instances, as in cases recorded by E. S. Reynolds, more than one member of a family has been affected. There is no undoubted instance on record, so far as the writer knows, in which a diagnosis of family disseminated sclerosis has been verified by autopsy. Some of the cases referred to have undoubtedly belonged to the category of the familial spastic paraplegias.

**Social Conditions.**—The disease is as common, possibly more so, among the fairly well-to-do upper and middle classes as among the lower grades of society. City life does not seem to predispose to it; indeed, the frequency with which the symptoms occur in otherwise healthy young adults who have lived a country life is rather striking.

<sup>1</sup> *Inaug. Diss.*, Leipzig.

<sup>2</sup> *Deutsche med. Wochenschr.*, 1903, No. 41.

**Emotion.**—Mental shock, fright, worry, or grief is sometimes blamed by the patient as the cause of the symptoms, which certainly appear to be aggravated by psychical disturbances.

The symptoms sometimes appear after fatigue or exposure to cold.

**Pathology.**—Disseminated sclerosis is characterized anatomically by plaques or islets of disease scattered haphazard throughout the central nervous system. The cranial nerves are often involved, while degeneration of the spinal nerve roots has been observed. A classification into cerebral, spinal, and cerebrospinal types, although of clinical value, cannot in the pathological sense be strictly adhered to, since it seldom if ever happens that the patches are confined to the brain or spinal cord.

The pia mater has been sometimes described as unduly adherent. A slight degree of atrophy of the cerebral convolutions may be occasionally observed. In one or two exceptional instances, as in a case examined by the writer, the unusually small size of the spinal cord attracted attention. Glistening patches of variable size and shape and of grayish color are in some cases to be seen shining through the pia mater which covers the surface of the cord, and on section these are seen to correspond with diseased plaques situated in its substance.

Upon section, islets of a gray or grayish-pink color are, as a rule, readily recognizable. It may be, however, that the diseased patches cannot be detected until the tissue has been placed in Müller's fluid or chrome alum. A slight degree of shrinking of the cord is sometimes observed in the neighborhood of patches of old standing, yet the contour of the transverse section is seldom modified in a striking manner thereby. The islets of disease are often firmer than the surrounding tissue; indeed, they may have almost the consistence of india-rubber. Sometimes the plaques are distinctly softer than the normal cord.

The patches vary greatly in size and shape. In the brain they may be as large as a walnut; the majority, however, are very much smaller, and many are only recognizable on microscopic examination. The greater part of the transverse section of the spinal cord may be involved at one level, while a section a centimeter above or below may present an almost healthy appearance. A patch involving a large area may sometimes be traced through several segments. The patches may be wedge-shaped, rounded, oval, or quite irregular in contour; indeed, there is no characteristic shape. It is most unusual to find a patch of any size which does not extend to the periphery of the cord. The one situation in which the writer has found a notable exception to this rule is in the posterior columns. In two cases a large oval patch extended through several segments in this region.

It has been affirmed that certain parts of the transverse section are more liable to suffer than others. Thus most observers hold, and with this conclusion the writer is inclined to agree, that the white matter is especially implicated. E. W. Taylor,<sup>1</sup> from a study of eight cases, concludes that both gray and white matter are irrespectively involved. Ribbert asserts that the gray matter often offers a barrier to the spread of the diseased

<sup>1</sup> *Journal of Nervous and Mental Disease*, June, 1906.

area. The writer's personal observations are in accord with those of Dejerine and E. W. Taylor, who failed to find evidence in support of this conclusion. An examination of seven cases lends no support to Obersteiner's statement that the lumbar region of the spinal cord is less commonly involved than the parts above this level.

Bourneville and Guerard regarded the long tracts, Ziegler and E. Müller the posterior columns, as the regions of predilection. Obersteiner and others have noted a striking tendency to symmetrical affection of the spinal cord, medulla, and pons. This is, however, far from a constant feature. Williamson and Rossolimo have reported cases in which the distribution of the patches corresponded to that of the bloodvessels. In not one of the seven cases examined by the writer was it possible to assert that this was so.

In the brain the basal ganglia and the neighborhood of the ventricles, the corpus callosum, the pons (Strümpell), centrum ovale (Gowers), and the neighborhood of the olives are favored sites. For long it was supposed that the cerebral cortex was always spared. Oppenheim, E. W. Taylor, and Sander have shown that patches in the cortex are by no means uncommon. Islets of disease are comparatively rarely met with in the cerebellum. Weigert, however, has drawn attention to diffuse changes consisting in an increase of the tangential and radial fibres in this situation.

For long it has been known that patches of sclerosis may occur in all the cranial nerves in some part of their course. The optic nerve and tract appear to be favorite situations. Changes in the anterior and posterior spinal nerve roots have been noted. On the other hand, it is doubtful whether the peripheral spinal nerves are ever implicated, although it must be admitted they appear to have been seldom systematically examined.

**Histology.**—The two most striking features of the diseased areas are the disappearance of the myelin sheaths and the increase in the neuroglial tissue. The consistence of the patches varies with the degree of neuroglial proliferation. Tredgold<sup>1</sup> divides the islets of disease into three varieties, hard, soft, and intermediate. Hard islets occur chiefly in the cord, and consist of a dense interlacement of thickened neuroglial fibres with very few glia cells. Nerve fibres are practically absent, although occasionally a naked axis cylinder is met with. There are no products of degeneration and but few bloodvessels. Soft islets were confined to the brain in Tredgold's three cases, usually have a well-defined margin, often surrounded by a zone of leukocytes, and consist of a loose reticulum containing a semi-fluid material. They are generally devoid of nerve cells or fibres, while there is no neuroglial proliferation, and usually no degeneration products. The vessels are often much engorged. Intermediate islets contain nerve fibres in every stage of degeneration. Products of degeneration are abundant, while the neuroglia is somewhat thickened and has a finely granular structure. Glia cells are often plentiful, the vessels are distended, and their lymphatics filled with leukocytes and fat-containing cells. Although the borders of the patches usually appear well defined to

<sup>1</sup> *Rev. of Neural. and Psychiat.*, 1904.

the naked eye, under the microscope a gradual transition from diseased to healthy tissue is generally recognizable.

The changes in the myelin sheaths are well seen in the intermediate islets, particularly toward their periphery. The first alteration appears to consist in swelling of the myelin substance. Later the myelin sheaths begin to disintegrate, undergoing a process of lecitholysis or fatty degeneration. Finally, the myelin sheath disappears.

The axis cylinders persist after their medullary sheaths have disappeared. The unclothed axis cylinders do not, however, always present a healthy appearance. Some of them are swollen, many, on the other hand, are considerably reduced in diameter, and on longitudinal section are found to be fusiform in shape. Popoff, Erben, and Sträuber assert that these fine axis cylinders are newly formed, while Weigert and others believe that they are atrophic fibres. Bielschowsky traced the axis cylinder through a diseased patch to its continuation once more as a myelinated fibre. Kaplan demonstrated what he regards as a cement substance (axostrome) in the axis cylinder. Some naked axis cylinders retain their axostrome, others lose it.

Secondary degenerations, although described (Babinski, Jolly, Rossolimo), are undoubtedly uncommon, as the comparative integrity of the axis cylinders within the plaques might lead one to expect. Even when present they rarely extend more than a short distance from the plaques which produce them. The nerve cells of the spinal cord and posterior ganglia often show a degree of chromatolysis and an increase of the intracellular pigment, but these alterations are usually surprisingly slight.

The neuroglia in the firmer patches consists of a dense felt-work of fibres with few nuclei. Recent patches show a more open network.

Changes in the bloodvessels were first described by Rindfleisch. Ribbert found a large vessel situated more or less in the centre of most of the patches in those cases he examined, and in two instances he found appearances suggesting with great probability an arterial thrombosis. Dejerine and Thomas point out that the vessels may be more numerous than normal in the sclerosed areas. Thickening of the vessel walls has been described by several observers, and narrowing of the lumen with, indeed, actual occlusion has been recorded. Hyaline degeneration of the wall has also been noted. In a number of cases which have been examined no pathological changes in the bloodvessels have been met with (Taylor, Schüler, Buchwald, Jolly, the writer), while the circumstance that these changes when present are limited to the patches, and may well be secondary to the neuroglial increase, must be kept in view.

Rossolimo indicates that slight inflammatory changes are to be found in some cases in the neighborhood of the vessels. Thus the perivascular spaces are dilated and contain both mononuclear and polynuclear leucocytes in various stages of fatty degeneration. Borst, who also emphasized these changes in the perivascular lymph, holds that a lymph stasis may be a factor of importance.

**Pathogenesis.**—This is most obscure. The problem involves the nature and sequence of the histological changes and their cause. In the first place, the author sides with E. Müller, who, following Ziegler and Schmaus,

speaks of a primary as opposed to a secondary disseminated sclerosis, thereby excluding such cases as those described by Williamson, Marburg, and others, in which prominent changes in the bloodvessels and the distribution of the diseased areas suggest their dependence upon interference with the vascular supply. It is obvious that the diseased process may begin either in the neuroglia or in the parenchyma, or that it may affect both tissues simultaneously to a greater or less degree. E. Müller is a strong supporter of the view that the initial change consists in a neuroglial proliferation, as a result of which the nerve elements are strangled. In favor of his opinion he states that the patches are especially prone to occur in regions where the neuroglia is normally most dense, and, further, that the density of the neuroglia in disseminated sclerosis is much in excess of that met with in any other affection of the nervous system. Again, he points out that toward the edge of a plaque increase in the neuroglia may sometimes be seen surrounding perfectly healthy nerve fibres. Even if it be granted that the neuroglial increase is greatest in the regions in which the neuroglia is normally most dense, this is not a weighty argument in favor of the primary affection of the neuroglia. In cases of sharply-defined secondary degeneration in the spinal cord, a neuroglial proliferation which must be secondary to the atrophy of the nerve fibres is sometimes observed beyond the limits of the degenerated tract (Mott and others).

Very strong evidence against the primary neuroglial origin of the disease is afforded by the facts that in some patches very little neuroglial increase is to be observed, and that such islets may co-exist in the same case with others in which the neuroglial proliferation is excessive. Further, the rapidity with which symptoms develop and the manner in which they vary in intensity are difficult to explain on this hypothesis.

The view, which is supported among others by Risien Russell,<sup>1</sup> and Tredgold, that the primary change is in the parenchyma, the neuroglial increase being a compensatory process, has much more to commend it in the opinion of the writer. The strongest argument in support of this view is the circumstance that very profound changes are sometimes found in the myelin sheaths, with little or no increase of the neuroglia in the same region. Toward the periphery of a patch, for instance, degenerative changes are sometimes found in nerve fibres which are lying in the midst of perfectly healthy neuroglia.

A third body of observers takes up the position that both the myelin sheaths and the neuroglia suffer primarily as a result of a common cause. Erb, Gowers, Rossolimo, and others regard the process as a form of chronic myelitis. E. Müller and E. W. Taylor stoutly deny any evidence of inflammatory changes in the very considerable material at their disposal. Strümpell, who holds very similar views to Müller, regards the condition as a multiple gliosis predisposed to by a congenital factor, although determined, it may be, by a variety of exogenous causes.

Ribbert, E. W. Taylor, Strähuber, T. Buzzard, and others believe that some toxin of undetermined nature circulating in the blood is the agent responsible for the histological changes. Alexander Bruce believed that

<sup>1</sup> *Allbutt's System of Medicine*, 1899, vii, 83.

the plaques were caused by a gradual infiltration of the tissues by the toxic lymph spreading from a central focus until it exhausts itself.<sup>1</sup> W. E. Bulloch<sup>2</sup> has recently succeeded in producing paralysis of the limbs in rabbits injected subcutaneously with the cerebrospinal fluid from a case of disseminated sclerosis. On histological examination vascular engorgement and fragmentation of the myelin sheaths were observed and in the late stages degeneration throughout the cord.

**Symptoms.**—As the multiplicity of the lesions and their widespread distribution would lead one to expect, the symptoms are numerous and varied. A classification into three clinical types—spinal, cerebral, and cerebrospinal—as suggested by Charcot, can only be accepted in a relative sense, for cases are seldom met with in which the symptoms are exclusively spinal, while still more rarely is a diagnosis made in the presence of purely cerebral manifestations. The circumstances, first, that the patches are scattered indiscriminately throughout the nervous system, and secondly, that in the spinal cord the motor fibres are gathered into small compass, readily explain the frequency with which paresis or paralysis of the lower limbs is observed in the early stages. Moreover, small patches scattered through the brain substance, unless they implicate special situations, may fail to afford indications of their presence.

The characteristic features of the morbid process, viz., the preservation of the axis cylinders and ganglion cells, and the destruction of their medullary sheaths, may account for the special characters of certain symptoms, such as the peculiar form of optic atrophy, the absence of muscular wasting and the frequency of fleeting symptoms.

**Motor Symptoms.**—Weakness in the lower extremities is almost always complained of, even in the earliest stages. The paresis may be so slight that its presence is only perceptible after prolonged exercise. Thus, undue fatigue in one or both lower extremities, after a long walk, is one of the commonest early symptoms. The loss of power in the limbs, which is often asymmetrical, may be so pronounced that it is only with difficulty that the patient can walk up stairs or across the room. In advanced cases the legs may be completely paralyzed. Temporary paralysis of one arm is by no means uncommon as an early symptom. A slight degree of weakness in the hands and arms is often present, yet persistent paralysis in the upper extremities is exceptional unless in the terminal stages. Pronounced muscular atrophy is very rare. When this symptom is present the case may simulate amyotrophic lateral sclerosis.

Muscular hypertonicity is usually associated with the paraplegic state. The spasticity is slight in some cases; occasionally it is so pronounced that this rather than the motor weakness is the chief cause of difficulty in locomotion. Sudden flexor spasms often constitute a troublesome symptom, since they interfere with sleep. In the later stages of the disease flexor contractures tend to develop.

A slight degree of ataxia is a common early symptom in the upper limbs. The uncertainty of the required movement is clearly to be distinguished from the intention tremor presently to be referred to. Ataxia in the lower

<sup>1</sup> *Rev. of Neurol. and Psychiat.*, 1911, ix.

<sup>2</sup> *Lancet*, Oct. 25, 1913.

extremities may usually be demonstrated; indeed, in some cases this symptom constitutes the most striking feature of the picture, even in the earliest stages. The ataxia is, as a rule, of the cerebellar type, and hence is little increased when the eyes are closed.

The *gait* is usually altered at a very early date, yet it is sometimes surprising to find no appreciable alteration even in the presence of certain evidence of pyramidal disease. A common type of progression is the spastic paretic gait, in which there is a combination of spasticity and weakness, one leg being often more affected than its fellow. The patient shuffles along, dragging his feet as if the toes were stuck to the ground. In addition to the spasticity there is often some unsteadiness in walking, a condition, in short, of ataxic paraplegia. Sometimes, indeed, incoordination is the conspicuous feature; the ataxia, however, differs notably from that of tabes, for it is rarely increased when the eyes are closed. The stiff way in which the patient holds his head and the rhythmic tremor in head and trunk met with in the classical type are almost pathognomonic. The gait is sometimes staggering and reeling, as in cerebellar disease.

Another symptom, and one which is so characteristic that Charcot included it with nystagmus and speech defect as one of the cardinal signs of the disease, is the so-called *intention tremor*. So long as the patient is in a posture of complete muscular relaxation no tremor is to be observed; if he raises his hand with the object of carrying out some definite movement, the tremor commences. It is well brought out on attempts to write. Even when the patient is able to touch the point of the nose with the finger tip without any tremor, if the finger is kept in contact with the nose for a few seconds a rhythmical tremor of small amplitude may manifest itself. A test often employed in eliciting this symptom is to ask the patient to lift a glass of water to his mouth when the muscular effort required, together with the associated mental perturbation, commonly intensifies tremors. The rapidity of the tremor varies from four to six per second. The tremor may spread to other groups of muscles not directly called into action by the required movement. The intention tremor is usually most marked in the hands, but it is also seen in the lower extremities. It may be seen also in the head and neck during the act of walking. This constitutes no exception to the statement that the tremor only occurs during the performance of a purposive act, for while the patient is sitting or walking the muscles of the neck require to be constantly in a state of contraction to maintain the poise of the head. Tremor of the face and jaw has occasionally been observed. Intention tremor, when present, is one of the most characteristic symptoms, yet it is often absent.

Much discussion has taken place as to the origin of the tremor. Charcot was of opinion that it was due to an irregularity of conduction of the motor impulses through the sclerosed areas, a result of defective insulation of the axis cylinders or changes in them consequent upon disease of the medullary sheaths. Others hold that it is due to disease located in a particular situation, and in support of their view, point out that intention tremor is absent in cases in which the disease is practically limited to the spinal cord.

**Sensory Symptoms.**—The older writers emphasized the absence of sensory disturbances. This experience is quite contrary, that of more recent



observers, by whom subjective sensory symptoms, often of a fleeting character, it is true, are regarded as among the most characteristic of the early signs. Feelings of numbness, formication, deadness, tingling and pins and needles, particularly in one or both hands or feet, are very common. A girdle sensation is of comparatively frequent occurrence, while a sensation of constriction around a limb is occasionally experienced. Shooting pains, especially in the lower limbs, are sometimes complained of. Giddiness, a common symptom, may occur early, and often it occurs in paroxysms. Headache, although seldom severe, is often present. Vomiting is an occasional symptom, and may occur in association with paroxysms of headache or dizziness, or independently. A fixed pain in the back situated over the lower part of the sacrum is of frequent occurrence. Objective sensory defects of slight degree may be detected in most cases at one time or another if carefully looked for. Thus, Freund, who made a special study of 33 cases, found some definite alteration in no less than 29. All forms of sensation may be implicated. The hands and feet are most often involved, but hemianesthesia is not uncommon. The very slight degree of the objective sensory disturbances and their fleeting nature are very characteristic.

**Tendon-jerks and Skin Reflexes.**—The knee-jerks are almost always exaggerated and the increase is often more marked on one side than the other. In some cases a patella clonus can be elicited. Absence of the knee-jerks has been recorded, but is undoubtedly very rare. The writer cannot recall ever having seen such a case. The activity of the tendo-Achilles jerk is almost always increased. Ankle clonus, usually bilateral, but sometimes confined to one side, is demonstrable in a considerable proportion of cases. The tendon reflexes in the upper extremities are, as a rule, abnormally active.

The great value of Babinski's sign or the extensor plantar reflex as an indication of disease of the corticospinal descending tract has been amply confirmed. The collected experience of recent observers shows that in disseminated sclerosis, even at the earliest date at which a diagnosis is possible, the plantar reflex is almost always of the extensor type. Moreover in the writer's experience Babinski's sign is usually present in its most typical form, the slow dorsiflexion of the great toe being easily elicited from the whole plantar surface. A crossed plantar reflex may sometimes be demonstrated (Byrom Bramwell).

The *abdominal reflexes* have attracted considerable attention in this malady. Observations indicate (1) that their absence is specially characteristic of disseminated sclerosis, and (2) that they disappear early in the disease. Thus, Strümpell, who examined 185 persons free from nervous disease, noted their absence in 13.5 per cent. of these; while among 24 cases of disseminated sclerosis they were absent in 67 per cent. E. Müller found that the reflex was only absent in 5 per cent. of individuals free from nervous disease, as compared with 62.5 per cent. in 47 cases of disseminated sclerosis. The latter writer remarks that although the abdominal reflexes fail in many organic affections of the nervous system, they never fail with so much regularity as in disseminated sclerosis. There is nothing of special importance to note with regard to the cremasteric and other superficial reflexes in this disease.

**Micturition and Defecation.**—Disorders of micturition are ultimately present in all cases. Notwithstanding contrary statements, evidence of disturbance of the function of the bladder, often so slight as to cause little or no inconvenience to the patient, is usually forthcoming if carefully inquired for even before the more prominent symptoms are fully established. Thus in 80 per cent. of Oppenheim's cases there was some disorder of micturition. The writer obtained a history of some disturbance in 30 out of 34 consecutive cases in which a special inquiry was made. "Precipitate micturition," or a feeling of inability to retain the urine immediately the desire to empty the bladder is felt, is the abnormality usually first experienced. Another indication of defective function which usually appears at a later date is the delay which precedes the commencement of the act. The patient may require to exert considerable pressure before the urine begins to flow. When the urine commences to flow it comes in a stream of full volume, although it may be defective in force. Retention of urine, a not uncommon symptom later, is a complication which must always be regarded with anxiety on account of the liability to cystitis. Incontinence is common in advanced cases, and is usually of the remittent type, but rarely depending upon complete paralysis of the bladder wall. Like so many of the symptoms of this disease, the bladder symptoms often vary from time to time.

Constipation is the rule. Some loss of control is also often seen when any tendency to looseness of the bowels exists. True incontinence of feces is exceptional unless in the later stages.

The sexual functions are at times interfered with. Impairment both of sexual desire and power has been described, and an exaggerated sexual desire has been noted in a few cases.

**Defects of Vision and Optic Atrophy.**—*Visual defects*, which are frequent and characteristic, often appear at an early date. A temporary unilateral amblyopia of sudden onset and often regarded as hysterical in origin, since it is unaccompanied by changes in the fundus oculi, may indeed be the first indication of disease. *Diplopia* with no visible impairment of the ocular movements, another early symptom for which the patient seeks advice, was present in 13 of 31 cases recently examined by the writer. Although the visual defects are often fleeting, a permanent diminution in central vision, associated with atrophic changes in the optic disks, is often met with. Unlike the amblyopia of tabes, this defect of sight very rarely progresses to complete blindness. A central scotoma, usually bilateral, it may be for color alone, is often to be found if carefully looked for, and when present is a valuable aid in diagnosis. Concentric contraction of the field of vision is found in some cases. Uhtoff,<sup>1</sup> who has studied the alterations of the visual fields in 150 cases, describes six groups of cases: (1) In one-half of all cases with anomalies of the visual fields, central scotomata, usually bilateral, often relative, with no alteration of the periphery of the field, are present. (2) In rare cases peripheral contraction of the field occurs together with scotomata. (3) In a considerable percentage of cases there is an irregular peripheral contraction of the field. (4) Isolated

<sup>1</sup> *The Ophthalmoscope*, 1905, p. 429.

cases present regular concentric contraction. This condition occurs in pure multiple sclerosis as well as in those cases which are complicated by hysteria. (5) In one case there was a ring scotoma, and in another (6) a central scotoma was present, but cleared up, and then a persistent contraction of the field appeared.

Atrophic changes in the optic disks are of very great importance. There may be some pallor of the temporal side of the disk, or an incomplete pallor of the whole papilla, so that the inner part still retains a pinkish tint, or, when the process is more advanced, the whole papilla may be uniformly pale. As T. Buzzard<sup>1</sup> pointed out, slight differences in the color of the papillæ on opposite sides is often of the greatest value in determining whether the appearance is to be regarded as pathological. The frequency of recognizable pallor of the disk has been variously estimated. Uhtoff found that the papillæ presented a pathological degree of pallor in 37 per cent. of all cases, and other observers have found an even higher percentage. The dependence of the changes in the optic disks upon an islet of disease situated in the optic disks has been repeatedly demonstrated.

A small number of cases have been observed in which there was a slight degree of optic neuritis, never amounting to a true papillitis. The appearances, which are those of a retrobulbar neuritis, have been proved to be due to the presence of a plaque in the immediate vicinity of the papilla.

**Ocular Muscles, Nystagmus, Pupils, etc.**—The ocular muscles are particularly liable to be affected (Uhtoff, 20 per cent.). Diplopia has been mentioned. Usually when this symptom is present the paresis is so slight in degree as to escape detection unless specially examined for by the appropriate visual tests. The sixth nerve is most commonly affected. Paresis of the third is usually partial. These muscular defects are almost always of slight degree, but complete ophthalmoplegia has been recorded. Ptosis either confined to one side or bilateral is not infrequent. Wildbrand and Sanger lay stress on this symptom, believing that in certain cases it may be of even greater diagnostic importance than nystagmoid movements. Müller, who met with ptosis in 8 per cent. of his cases, also observed paresis of lateral conjugate movement in three and of upward movement in two cases.

*Nystagmus* belongs to the symptomatic triad formulated by Charcot. Three degrees may be conveniently recognized: (1) Nystagmus which occurs while the eyes are in a position of rest; (2) nystagmus which is only present in association with a fixed purposive movement, such as lateral or vertical deviation or upon fixing a near object; and (3) movements which occur during fixation, but which rapidly cease and are at the same time frequently irregular both in time and amplitude. To this last group we would reserve the term nystagmoid movements. In few cases of disseminated sclerosis do we find the type which we have classed in the first group. On the other hand, the second and third types are very common. The movements are usually fairly rapid and regular, but vary considerably in amplitude in different cases. Coarse nystagmus, for example, may be elicited when the patient looks to one side, while when he looks in the oppo-

<sup>1</sup> *British Medical Journal*, 1893, ii, 779.

site direction the movements may be much finer in quality. The nystagmus is usually most obvious when the patient looks to the extreme left or right and at the same time directs the eyes slightly upward; sometimes, however, when he looks directly upward or downward the movements are also visible. Occasionally the movements are only to be seen when the eyes are fixed at the extreme of the vertical plane. The direction of the oscillation is commonly from side to side, the two eyes acting in unison. A certain amount of rotation of the eyeball is by no means uncommon, and in rare cases the oscillations take place in a vertical direction. The writer has seen one case in which the movements were confined to one eye.

Kunn has noted a "Zitterbewegung" in the ciliary muscle. The writer has also observed evidence of rhythmic contractions in this muscle on ophthalmoscopic examination. Alterations in the size and activity of the pupil are not uncommon. There was a difference in the size of the pupils in 24 per cent. of Uhtoff's cases. Parinaud remarks that inequality of the pupils occurs early in the disease, and that miosis when present is spastic as opposed to the paralytic miosis of tabes.

The pupils react well on convergence provided that the movement of convergence is not itself defective. The light reflex, although sometimes sluggish, is very rarely lost. Müller notes that among 364 cases observed by Fränkl-Hochwart, Probst and Uhtoff, the Argyll-Robertson pupil was only present in 4. Fränkl-Hochwart met with hippus in 12 cases.

**Speech and Articulation.**—In the older description of this disease speech affections occupy a prominent position. They are, however, by no means of constant occurrence. Byrom Bramwell found some defect of speech in 62 per cent. of his series, while some alteration was present in two-fifths of Müller's cases. Among 33 consecutive cases recently examined by the writer, in 15 (45 per cent.) speech was altered.

It may be that the patient notices some slight difficulty in speech, especially toward the end of a sentence, even before any change can be detected by the observer. The alteration in speech in the early stages of the disease may be so slight that it is difficult to describe, and yet it may be sufficiently characteristic to the experienced observer to help him greatly in making his diagnosis. The slow and measured way in which the patient enunciates and a singular monotony of voice are usually the salient features. When he reads aloud it is interesting to note the way in which he tries to vary the pitch of his voice, of the monotony of which he is obviously conscious. At a later date he chops his words, each syllable is pronounced slowly, hesitatingly, distinctly, and precisely. This is the syllabic, scanning, or staccato speech which has been compared by Charcot to that of incipient intoxication, by Oppenheim to that of a child learning to spell, and by Marie to that of an individual who is steadily but surely ascending a hill. Each syllable is produced with apparent effort, and this is accentuated by the associated tremor often present. Müller found a true staccato speech in only 15 per cent. of his cases. Certain letters, notably l, p, g, b, and d, are especially apt to give rise to difficulty in pronunciation. Later in the course of the disease, defective articulation may render speech quite unintelligible. As Oppenheim observes, when an attempt is made to intone the letter e, a distinct tremor in the voice is often

noticeable. This observer has drawn attention to the scanning character of the respiration, to which he is inclined to attribute the corresponding peculiarity in speech. Goldscheider believes that the monotonous character of the speech may be due to defective control over the act of expiration, for he has shown that the augmentations and interruptions in the expiratory blast, which he has demonstrated take place during the act of speaking, are in this disease abnormally slow and defective. Aphonia is occasionally present. Réthi has given a detailed description of the appearances in 44 collected cases in which the larynx was examined. In 63 per cent. some form of laryngeal paralysis was present, while in half the cases tremor of the vocal cords was observed.

**Mental Symptoms.**—It is somewhat surprising to find that profound psychic alterations are exceptional. Even in fully developed cases there may be no obvious mental disturbance. As a rule, however, after the disease has been in progress for any length of time, a certain degree of mental impairment is noticeable. Thus in Byrom Bramwell's series of cases defective memory was present in 50 per cent., some emotional alteration in 40 per cent., and further evidence of intellectual impairment in 27 per cent., while E. Müller found a slight enfeeblement of the psychical faculties in about 25 per cent. The psychic alteration shows itself especially in enfeeblement of memory, in a certain slowness of thought, in the apathy and indifference with which the patient regards his surroundings, and in a degree of stupidity which the facial expression would almost lead one to expect. A sense of well-being, or euphoria akin to the *spes phthisica*, is often observed, less commonly a depression of spirits which has been known to pass into actual melancholia. Emotional control is often impaired. The occurrence of outbreaks of emotion does not necessarily imply a corresponding defect in the intellectual capacity. It is interesting to note that several cases have been reported in which a pathological examination established the co-existence of general paralysis of the insane.<sup>1</sup>

**Trophic and Vasomotor Changes.**—These are extremely rare and of corresponding minor significance. James Collier has reported five cases in which he noted the presence of erythromelalgia. There are a number of cases on record in which muscular atrophy was present, although a pronounced degree of wasting is altogether exceptional. Bedsores may develop in the terminal phases.

**Congestive and Epileptic Attacks.**—Attacks similar in character to the congestive attacks of general paralysis of the insane have been recorded. Their rarity may be gathered from the fact that the writer, who has examined over 150 cases of this disease, can only recall one instance. Fränkl-Hochwart met with apoplectic attacks in 10 of his series. The attacks have been sometimes preceded by giddiness, the patient falls to the ground, is sometimes convulsed, and may remain comatose for several hours. The temperature is raised. Hemiplegia is an occasional sequel. Sudden attacks of giddiness and faintness without loss of consciousness are less uncommon. Müller has met with two cases in which Jacksonian attacks occurred. Epileptic attacks were noted in 3 of Fränkl-Hochwart's 206 cases.

<sup>1</sup> Hunt, *Am. Jour. Med. Sci.*, 1903, cxxvi, 126.

**Earliest Symptoms.**—Motor weakness in the limbs, and especially in both lower extremities, paresthesias in the limbs, giddiness, tremor or ataxia, and amblyopia are, in the order stated, the most common early symptoms. The development of the initial symptoms as complained of by the patient has been studied by Byrom Bramwell (110 cases), who states: "In 41 cases the initial symptoms occurred in the extremities, and were purely motor or ataxic; in 3 cases the symptoms were purely sensory; in 22 cases the symptoms were both sensory and motor; in 24 cases the symptoms were purely cerebral or ocular; in 17 cases there were different combinations of motor and sensory symptoms with cerebral or ocular symptoms." Ashley W. Mackintosh, who has tabulated 80 cases, for the most part drawn from Ferrier's material, remarks on the extraordinary variability of the modes of onset, the comparatively large number of cases with acute or sudden onset (10 cases), the frequent occurrence of purely unilateral symptoms at the onset (15 cases), and the comparative frequency of the occurrence of sensory symptoms at the onset, either alone (10 cases) or combined with other symptoms (19 cases).

**Types.**—A number of clinical types are recognizable according to the preponderance of certain symptoms or different modes of onset.

1. The *classical* type, characterized by intention tremor, nystagmus, speech affection, and it may be other cerebral manifestations in association with a spastic or ataxic paraplegia.

2. The *cerebrospinal* type, in which even in the absence of the symptomatic triad of Charcot, symptoms alike cerebral and spinal point to widespread disease of the central nervous system.

3. The *hysterical* or *intermittent* type, in which fleeting symptoms suggestive of hysteria constitute the most striking feature of the case.

4. The *spinal* type, characterized by prominent spinal symptoms resembling a variety of spinal diseases. Of these, a progressive paraplegia unaccompanied by sensory disturbance and indistinguishable at first, it may be, from primary lateral sclerosis is very common. Again, a spastic paraplegia may develop with the rapidity of an acute myelitis. The co-existence of ataxia and paraplegia may constitute a symptom complex which closely resembles the combined systemic disease.

5. The *cerebral* type is much less common. Occasionally vomiting, giddiness, and headache are the most striking symptoms. In rare cases, a cerebral hemiplegia is met with as the leading feature of the case. Very exceptionally, bulbar manifestations or mental symptoms are the most conspicuous alterations.

6. The *optic* or *ocular* type in which some disturbance of vision or ocular palsy constitutes the prominent symptom.

7. The *sacral* type, recently described by Oppenheim and Kurt Mendel, in which pains in the legs, disturbance of the sphincters and sexual functions, point to a lesion in the lower part of the spinal cord.

**Diagnosis.**—This is not as a rule so difficult as the irregular distribution of the pathological lesions and the multiplicity of types might lead one to expect. At the same time there are few diseases which present such puzzling diagnostic problems. The diagnosis is based upon (1) the detection of certain characteristic symptoms or association of symptoms, upon

which alone a positive opinion may sometimes be advanced; (2) a history of variation in the severity of the symptoms, which, when present, is in itself so suggestive as to be almost pathognomonic; and (3) the exclusion of a variety of diseases, which may closely simulate disseminated sclerosis. The classical type presents features which are distinctive, for intention tremor, scanning speech, and nystagmus in association with a spastic or ataxic paraplegia and an extensor response place the problem beyond all doubt. These cases, however, form only a small percentage.

It is especially in the early stages that errors are made, notably when temporary manifestations disappear, leaving the patient in perfect health, or when the symptoms are such that the supposition of a single focus of disease appears to satisfactorily explain them. Weakness in the lower extremities, it matters not how slight, in a young adult, if accompanied by exaggerated tendon-jerks and Babinski's sign, should suggest to the physician the possibility of this disease, particularly when on examination no evidence of vertebral disease, no muscular wasting, and no objective sensory disturbances are detected. If in addition a degree of ataxia is present and the abdominal reflexes are absent, the suspicion is considerably strengthened. Further, if there is a history of previous temporary paresis or paresthesia in a limb or of a fleeting amblyopia or diplopia, the suspicion amounts almost to a certainty. A slight degree of ataxia in the upper extremities, the occurrence of vertigo, and, above all, pathological pallor of one or both optic disks in conjunction with a spastic or ataxic paraplegia render the diagnosis almost certain, even in the absence of a history of variations in the clinical picture or of nystagmus, intention tremor, or characteristic alterations in speech.

*Hysteria*.—Of the many diseases for which disseminated sclerosis may be mistaken, this takes the first place. Disseminated sclerosis often occurs in young women, its early symptoms are often transient and completely recovered from, while hysterical stigmata are often met with in association with the organic affection. To Thomas Buzzard especially is due the credit of showing the frequency with which disseminated sclerosis is mistaken for hysteria. Transitory paresis or paresthesia in a limb, even though completely recovered from, symptoms which some years ago would have been diagnosed as certainly functional, are to be regarded with the gravest suspicion, since they are so often the precursors of the affection under consideration. Even although a hemianesthesia with involvement of the special senses is detected, the observer must remember that the recognition of hysteria does not exclude associated organic disease. Nystagmus, pallor of the optic disks, and Babinski's sign justify a positive diagnosis of organic disease, while incontinence of urine, a symptom which it is true does not usually occur until the later stages of the disease, carries with it the same significance.

Acute disseminated *myelitis* or encephalomyelitis presents a very similar picture; some believe that it is a more intense degree of the same process. The symptoms develop acutely, it may be with fever, and reach their height in a few days. There is muscular wasting. Variations in the intensity of the symptoms do not occur. Progress is in the direction of improvement, although this is very often limited.

*Subacute combined degeneration* of the spinal cord may simulate disseminated sclerosis very closely. The diagnosis is very important, for these cases often run a very rapid course. In both diseases ataxic paraplegia is often the most striking feature. Subacute combined degeneration usually develops rather later in life, and its symptoms once present show no tendency to disappear. Pallor of the optic disks is not met with, while pronounced anesthesia, beginning in the feet and progressing steadily upward, is the rule. The symptom complex of the later stages, viz., the flaccid palsy, often suddenly developed, with abolition of the tendon jerks and complete loss of sphincter control, places the diagnosis beyond all doubt.

*Cerebrospinal syphilis* occasionally closely resembles disseminated sclerosis. The peculiar pallor of the optic disks, the presence of a color scotoma, and absence of the abdominal reflexes speak strongly for disseminated sclerosis. Scanning speech and intention tremor do not occur in cerebrospinal syphilis. The Argyll-Robertson pupil is almost conclusive evidence of syphilis of the nervous system, corroborative evidence of which is afforded by the detection of a cerebrospinal lymphocytosis or a positive Wassermann reaction. Improvement under antisyphilitic remedies may help to confirm the diagnosis.

*General paralysis of the insane* has been known to cause trouble in diagnosis. The writer has met with one instance in which this was so. The more profound mental alterations, a history of syphilis, and the existence of the Argyll-Robertson pupil, a cerebrospinal lymphocytosis or positive Wassermann reaction are data of importance.

*Intracranial tumor* is a diagnosis sometimes made in the early stages of disseminated sclerosis. The writer has seen this error made on three occasions by eminent authorities, the subsequent progress of the case demonstrating its true nature. Giddiness with vomiting, unsteady gait, nystagmus, and tremor may all result from a patch of sclerosis situated in the cerebellar peduncles. Headache may be present, though rarely severe, while in very exceptional cases there may be a slight degree of optic neuritis. The intensity of the symptoms due to increased intracranial pressure in most cases of new growth and the absence of remissions are points of importance in relation to the diagnosis.

*All forms of spastic paraplegia* in which the paralysis of the lower limbs is the dominant feature may cause difficulty. Many of the cases formerly classed as primary lateral sclerosis, an affection which we now know to be one of the rarest of organic nervous diseases, were without doubt examples of disseminated sclerosis. It may be, indeed, that there is at the present time a tendency for the pendulum to swing too far in the opposite direction. The possibility of amyotrophic lateral sclerosis must always be kept in view.

The differential diagnosis from *pseudo-sclerosis* and *diffuse sclerosis* will be considered under these diseases.

**Prognosis.**—It is very doubtful whether permanent recovery ever takes place although cases have been observed in which the patient has remained free from symptoms for several years. Thus, Oppenheim mentions three examples in which the patients had recovered and been in perfect health for from five to ten years, and similar instances have been met



with by others. No less uncommon, however, are cases such as those recently reported by T. Buzzard in which after intervals of several years the symptoms of the disease have reappeared. Time may show that very exceptionally permanent recovery may take place. For the present this question cannot be answered in the affirmative.

The disease tends in the majority of cases to run a *chronic course*, often interrupted, it is true, by remissions and relapses. There are instances in which death has occurred within a year, but these are exceptional. Byrom Bramwell has ascertained the duration of life from the appearance of the first symptom in the 35 fatal cases of his series. He found that in 10 cases (28.5 per cent.) death occurred in less than five years, in 23 (65.7 per cent.) in less than ten years, and in 31 (88.5 per cent.) in less than fifteen years; one patient died within a year. In no instance was the duration more than twenty-one years. On the other hand, among 61 non-fatal cases, 5 had lived at the time of writing for periods varying from twenty-two to thirty-three years. The average duration of life in 96 fatal and non-fatal cases was found to be ten and a half years.

There are practically no available data which permit of an opinion as to the probable course or duration of a particular type of case when seen in its early stages, although if bulbar symptoms or pronounced bladder trouble appear early the expectation of life will probably be less than the average.

**Treatment.**—Conclusions as to the beneficial effect of this or that line of treatment are naturally extremely difficult to arrive at in the case of an affection in which there is a tendency for intermissions and remissions to occur independently of all therapeutic intervention. No known remedy has been proved beyond doubt to exert a favorable effect, hence treatment is limited (1) to certain general directions indicated by common-sense; (2) the avoidance of circumstances which may be expected or have actually been proved to have a deleterious influence; and (3) to attempts to alleviate individual symptoms as they arise.

The spirit of optimism which these patients not uncommonly present should, so far as possible, be left undisturbed for even although it is perhaps unreasonable to suppose that such a mental state can favorably influence the disease, yet it is certain that prospects of improvement do much to make the patient's outlook on life more bearable.

Strict injunctions are to be laid down as to the avoidance of muscular exhaustion and in particular the patient should be cautioned when walking always to stop short of fatigue. Confinement to bed is, on the other hand, a mistake; indeed, the patient should rather be encouraged to regularly exercise his muscles. A warm climate may be recommended to those who can afford it. Plenty of fresh air and a nutritious diet are advisable. Worries of all kinds are to be, as far as possible, avoided. Hot baths are contra-indicated, experience having shown that sometimes harm is done thereby. Marriage should be forbidden. The prejudicial effect of pregnancy is undoubted and should be explained to female patients. The induction of abortion may be even justifiable, when it is ascertained that during a previous pregnancy the symptoms were intensified.

Ataxia when pronounced is at times benefited by a course of coördinated exercises, although the brilliant results sometimes obtained in the treatment of tabes are not to be expected. Hydrobromate of hyoscyne in doses of  $\frac{1}{150}$  to  $\frac{1}{100}$  gr. may influence the tremor. Flexor spasms, often a troublesome symptom, since they interfere with sleep, are occasionally alleviated by five to ten grains of veronal given at night. When rigidity of the limbs is extreme the cautery applied to the back on either side of the spine may prove of service. Incontinence of urine, if due to overflow from a distended bladder, is to be treated by the passage of the catheter at regular intervals, while if it is of the intermittent variety, benefit may result from tincture of belladonna in 10 minim doses. Obstinate constipation is best relieved by a daily soap and water enema.

Associated hysterical symptoms should be treated by a confident prediction that they will disappear and by the methods of suggestion and persuasion commonly employed in the treatment of such manifestations.

Although no drug has been definitely proved to have any effect upon the course of the disease, the writer is inclined to believe that arsenic in moderate doses, 2 to 6 minims of the liquor three times a day, is sometimes of service. Strychnine is to be avoided, since it tends to increase the spastic state. Silver nitrate and potassium iodide have, it is affirmed by some writers, a universal beneficial effect. Fibrolysin has been employed but apparently without definite benefit.

**Pseudo-sclerosis.**—Westphal described under this name two cases diagnosed during life as disseminated sclerosis, in one of which the autopsy was entirely negative, while in the other the change found was a general increase in the consistency of the brain. Strümpell gave the first distinctive description of the symptom complex. Fränkl-Hochwart<sup>1</sup> has studied the cases reported up to 1903, 13 in number. Two important contributions have since appeared, one by Fickler<sup>2</sup> who reported two cases and reviewed the literature. The other is by Rebizzi,<sup>3</sup> who regards pseudo-sclerosis and diffuse sclerosis as the early and late stage of the same affection, for which he proposes the title of Westphal-Strümpell's disease.

**Etiology.**—Bäumlin's four patients were sisters, and Rebizzi has described cases occurring in father and daughter. A strong neuropathic heredity has been a striking feature in several instances. In no case has there been any definite relationship to trauma. Males and females are equally affected. The symptoms developed in 5 cases during the first decade, in 3 during the second, in 1 in the third, and in 4 in the fourth.

**Pathology.**—No changes were found postmortem in the majority of the cases examined. It should be mentioned, however, that in 6 cases a microscopic examination of the brain was not made, and that the majority of the observations predated the introduction of elective staining methods. The consistence of two of the brains examined is described as abnormally dense. A spinal leptomeningitis, slight in degree, was found by Bäumlin in one case. Strümpell met with a very slight degeneration in the cervical portion of the pyramidal tracts in both his cases, in one of which similar

<sup>1</sup> *Arch. aus dem Neurol. Institut an der Wiener Universität*, 1903, Heft 10, p. 1.

<sup>2</sup> *Deutsche med. Wochenschr.*, 1904, No. 51.

<sup>3</sup> *Riv. di patol. nerv. e ment.*, vol. x.

changes were also observed in the cervical portion of Gowers' tracts. Rebizzi and Fränkl-Hochwart remark that the morbid anatomy of pseudo-sclerosis cannot be strictly separated from that of diffuse sclerosis, for cases are on record which might be included in either group.

**Symptoms.**—Mental changes are the rule. A degree of apathy, peculiarities of disposition, irritability, and attacks of temper are among the alterations noted, while maniacal outbursts with hallucinations have been reported. Epileptic attacks were present in 6 of 13 cases collected by Fränkl-Hochwart. Some disorder of speech was observed in 9 cases, in 5 of which speech was described as scanning, while in 2 it was unintelligible. Facial paralysis has not been noted, although in 3 cases a defect in facial mimicry was remarked on. The optic disks were normal in every case, with the exception of one instance reported by Fickler. The special senses have always been normal, as were the pupils. Nystagmus was present in one case only, and once paralysis of the oculomotor nerve was observed. Dysphagia was present in one case. Giddiness was complained of by three patients, while headache, faints, apoplectiform attacks, and uncontrollable outbursts of laughter were each noted on two occasions.

**Weakness** of the extremities is described in two-thirds of the reported cases. Three times the paresis was hemiplegic in distribution; in one case all four extremities were involved. The gait is referred to as "clumsy, ataxic, or unsteady" in 5 cases, twice it is noted as spastic, and in two instances the patient was unable to walk. Tremor in the upper extremities was a constant symptom. In four cases it occurred during repose. It was of greater amplitude and less regular than in disseminated sclerosis. Three times the lower limbs were affected, the head was implicated in 3 cases, while twice the face and tongue were also involved. Contractures, sometimes temporary, sometimes permanent, were present in 9 cases.

**Pains** sometimes rather intense, were occasionally complained of. Three patients suffered from paresthesias of various kinds. In one of Westphal's cases hyperesthesia in different situations was noted at times together with some loss of sense of position.

The tendon *reflexes*, examined in 10 cases, were once found to be normal, while on nine occasions they were exaggerated, clonus being present in five. In Fickler's case the superficial reflexes were absent, otherwise no anomalies of the skin reflexes have been met with. In 2 cases there was incontinence of urine, while in 1 there was incontinence of feces. Transitory vasomotor disturbances were occasionally noted.

In 3 cases death occurred in from nine to ten years, in 3 from four to six years, and in 4 from one to two years. Fränkl-Hochwart's patient lived for fifty-seven years after the onset of symptoms. Pronounced intermissions were present in three cases, while in the records of several others there are indications of the same.

**Diagnosis.**—Pseudo-sclerosis bears a very close resemblance clinically to disseminated sclerosis, from which it has rarely been distinguished during life. The chief points of importance which distinguish pseudo-sclerosis are the frequency of mental changes, notably attacks of excitement and pronounced dementia; the wild character of the tremor, which sometimes occurs during rest, a very rare occurrence in disseminated sclerosis; the oc-

currence of epileptic attacks, which were present in half the recorded cases of pseudo-sclerosis, but very rare in disseminated sclerosis; the absence of changes in the optic disks: the comparative insignificance of the paretic symptoms in the lower limbs until the later stages, and the relative rarity of bladder symptoms. The development of the earliest symptoms during the first decade would be a strong point in favor of pseudo-sclerosis, as would a history of a similar malady affecting a relative.

**Diffuse Sclerosis.**—This rare affection, like pseudosclerosis, bears a close resemblance clinically to disseminated sclerosis.

**Etiology.**—It commences not uncommonly in childhood, sometimes not until the third or fourth decade. Heredity does not appear to be of etiological importance. There is a history of associated trauma in about a third of the reported cases. A relationship with alcohol, meningitis, and syphilis has been claimed in individual cases.

**Pathology.**—The most striking feature is the dense consistence of the brain, which has been described by individual writers as leathery, like India rubber, elastic, and cutting like cartilage. The cut section looks almost like ivory. Weiss points out that where there is much shrinking, hydrocephalus with ventricular dilatation may result.

**Symptoms.**—These resemble pseudo-sclerosis so closely that it will be unnecessary to do more than indicate the main resemblances and differences. In both the mental alterations occupy a prominent place, though in diffuse sclerosis dementia is more common and more profound. Speech disturbances occur in a large proportion of cases of diffuse sclerosis, and are also more marked than in pseudo-sclerosis. Tremor occurs in about two-thirds of the cases while paresis in the lower limbs with an unsteady gait are about as common as in pseudo-sclerosis, although, as a rule, more pronounced in degree. Aphasia, present in a third of the reported cases, is unknown in pseudo-sclerosis. Facial paresis has been described in a number of instances, and dysphagia, a symptom only once observed in pseudo-sclerosis, was present in half the reported cases. Giddiness and epileptic attacks occur about as frequently in the two conditions. Muscular atrophy has been described in a few cases of diffuse sclerosis, never in pseudo-sclerosis. Objective disturbances of sensation are much more frequent in diffuse sclerosis, occurring in almost half the cases; incontinence of urine and of feces is frequent. The average duration of life is from one to three or four years, and the course is steadily progressive.

**Diagnosis.**—The distinctive features from disseminated sclerosis are almost identical with those laid down in considering the differential diagnosis of pseudo-sclerosis from that disease.

**Tuberous or Tuberoze Sclerosis.**—Bourneville<sup>1</sup> described in 1880, under the title "*Sclerose tubereuse des circonvolutions cérébrales*," a condition characterized by overgrowth of the neuroglia in circumscribed islets which project from the surface of the cortex cerebri and from the walls of the lateral ventricles. Vogt has in several communications drawn attention to the clinical features. A. W. Campbell,<sup>2</sup> in 1905, analyzed 20 cases. J. S. Fowler and W. E. Carnegie Dickson, who in 1910

<sup>1</sup> *Archiv. de Neurol.*, 1880, i, 91.

<sup>2</sup> *Brain*, 1905, p. 382.

collected 29 cases, give an admirable summary of the literature.<sup>1</sup> Of these cases 20 were males and 9 females. There was a well marked neuropathic heredity in 12 out of 20 cases. Syphilis is not a cause. Epilepsy and mental defect are invariable. In only two cases was the mental defect described as moderate. The convulsions began in just one-half the cases during the first year. Some patients appear to have been normal up to the time of the first convulsion. Contractures and spastic paralysis were noted in six cases only, tremor in none. Congenital dislocation tumors are met with especially in the brain, kidney, heart, and skin. The diagnosis may be made "by the recognition of some of the associated tumors in a mentally defective person who is, or has been subject to epileptiform convulsions." Eight of the patients died by the fifth year and 20 by the sixteenth, while one patient lived to 35.

**Miliary Sclerosis.**—Under this name Gowers<sup>2</sup> described a case in which "throughout both hemispheres of the brain, and in all parts of them, the cortex contained minute reddish-gray spots at the junction of the gray and white substance." There was wasting of the nerve elements and an increase in the connective tissue in the affected areas. The patient was a man, aged fifty years; the chief symptom was general weakness of the limbs, with some rigidity and a few unilateral convulsive attacks. The speech became mumbling and unintelligible, and the patient became comatose and died. There was history of syphilis. The total duration of the symptoms was about ten weeks. Greiff has described similar appearances in general paralysis of the insane.

<sup>1</sup> *Quart. Jour. of Med.*, 1910-11, p. 43.

<sup>2</sup> *Lancet*, 1886, i, 145.

## CHAPTER V.

### THE DISEASES OF THE MENINGES.

By EDWIN BRAMWELL, M.D., F.R.C.P. (LOND. AND EDIN.).

**Anatomical and Physiological Considerations.**—The central nervous system is invested by three membranes: the dura mater, the arachnoid mater, and the pia mater. The dura mater, a dense fibrous tissue membrane, divisible into two layers and lined on its inner surface by endothelium, lies in close contact with the inner table of the skull and is continued as a fibrous sheath on to the cranial nerves. Three prolongations of the dura mater, the falx cerebri, tentorium cerebelli, and falx cerebelli, extend into the cranial cavity and play an important rôle as supporting structures. The dura mater is for the most part irrigated by the middle meningeal artery, while the trigeminus is its source of nerve supply.

The *arachnoid mater* and *pia mater*, the leptomeninges, are commonly described as the “pia arachnoid,” since disease processes which affect the one always involve the other. The pia mater is a thin vascular layer of fibrous tissue, which lies in the closest contact with the outer surface of the central nervous system. A prolongation of the pia mater, the velum interpositum, passes into the transverse fissure of the brain, carrying with it the choroid plexuses. The arachnoid, on the other hand, is a non-vascular fibrous tissue membrane. It lies external to the pia mater, with which it is connected by a delicate meshwork of fibrous tissue strands, and it is covered on its outer surface by a layer of endothelium. The arachnoid does not dip into the sulci as does the pia, but bridges them over. The inner surface of the ventricles is lined by a layer of endothelium known as the ependyma.

The subarachnoid space which lies between the pia and arachnoid contains the cerebrospinal fluid. Its meshes communicate freely with one another. The two membranes are widely separated where they lie between the cerebellum and medulla, and the space so formed is sometimes known as the posterior subarachnoid space or reservoir. The foramen of Magendie and the two adjacent foramina of Luschka offer the only means of communication between the subarachnoid space and the intraventricular system.

The *cerebrospinal fluid* is a perfectly clear fluid with a slight alkaline reaction and a specific gravity of 1006. Its principal constituent is sodium chloride. Traces of carbonates and phosphates are also present. It contains traces of globulin and of urea, also a reducing substance which is probably glucose. With the exception of a few lymphocytes it is devoid of cellular elements. Although it is held by some that the cerebrospinal fluid is a transudation, the researches of Halliburton as to its chemical composition afford strong evidence for believing that it

is not an exudation, but a true secretion. The fluid is in all probability secreted by the ependymal cells, which cover the choroid plexuses, and the secretion is probably continuous. The amount produced in the twenty-four hours has been variously estimated, some observers placing the quantity as high as one or two liters. Observations upon this point are possible when after certain cerebral operations the cerebrospinal fluid which escapes from the wound can be collected. It is, however, somewhat doubtful whether these results can be regarded as a true estimate of the normal secretion, since the operative procedure and the pathological condition for which it has been undertaken may alter its amount (Cushing). The fluid fills the whole intraventricular system, and through the foramina of Magendie and Luschka has free access to the cerebrospinal subarachnoid space. Since the secretion of fluid is in all probability continuous, it follows that in order to regulate the intracranial pressure there must be a constant return of the fluid into the blood stream. From the therapeutic standpoint it is of importance to remember that the nerve cells are surrounded by pericellular lymph spaces, that these are irrigated by cerebrospinal fluid, that medicinal substances before they can enter the cerebrospinal fluid have to be secreted by the secreting cells which cover the choroid plexuses of the brain, and that these cells have a selective action.

### PACHYMEINGITIS.

The diseases of the dura mater are naturally divisible into two groups, according as the pathological process is situated on the outer or inner surface of this membrane.

**Pachymeningitis Externa.**—Inflammation of the outer surface of the dura mater is uncommon. It occurs almost always as a secondary consequence of neighboring disease. A cerebral and a spinal form are described.

**Cerebral.**—The outer surface of the cerebral dura mater lies in close contact with the inner table of the skull, hence external pachymeningitis in this situation is always localized. It may be a sequel to fracture of the skull with extravasation of blood on the outer surface of the dura, erysipelas of the scalp, syphilitic caries, or tumors of the cranium. The extension of an inflammatory process arising in the labyrinth or mastoid cells may result in a localized external pachymeningitis or an extradural abscess. The clinical features may be overshadowed by those of the primary affection.

**Spinal.**—The dura mater which surrounds the spinal cord does not lie in contact with the bony canal, but is separated from it by a space, which is occupied by loose areolar tissue. Hence pachymeningitis externa spinalis may be either widespread or localized. In the great majority of cases the process is secondary to disease of adjacent structures. A bed sore perforating the sacrum may set up an acute and extensive perimeningitis; the infection may even penetrate the dura and lead to a leptomeningitis. Very rarely does a leptomeningitis extend through the dura and cause inflammation on its outer surface. Primary acute osteo-

myelitis or periostitis, actinomycosis, syphilitic disease of the vertebræ, or an aneurism which has eroded the vertebral bodies, may determine a localized external pachymeningitis. Tuberculous disease of the spine is a much more common cause. The dura which is often thickened is surrounded by a mass of tuberculous granulation tissue, in which caseous foci and abscesses of variable size may be contained. The tuberculous tissue is firmly adherent to the dura, the inner surface of which is usually smooth. The dura, however, may be infiltrated, and the morbid process may even extend through to its inner surface. In very rare cases tuberculous pachymeningitis is a primary affection (Schlesinger).

**Symptoms.**—These vary according to the nature and extent of the lesion upon which they depend. Thus when the process is acute and diffuse, pain in the back, root pains, muscular spasms, and hyperesthesia, with elevation of temperature, are met with. Again, when the pachymeningitis is localized and chronic, symptoms indicative of increasing pressure on the spinal cord, similar to those which occur in the case of a tumor, are produced. Indeed, in cases of tuberculous pachymeningitis in which there is no spinal curvature or evidence of disease of the vertebræ the diagnosis from tumor may be difficult if not impossible.

**Treatment.**—The treatment is that of the primary disease.

**Pachymeningitis Interna.**—A purulent internal pachymeningitis occurs as a very rare result of an extension from inflammation of the leptomeninges. Osler has met with a pseudo-membranous inflammation of the lining membrane of the dura as a secondary process in pneumonia.

**Pachymeningitis Interna Hemorrhagica.**—This condition is sometimes known as hematoma of the dura mater. It is customary to describe it with inflammatory diseases of the meninges, although it is not unlikely that the initial change is a hemorrhage from the inner surface of the dura.

**Pathology.**—This has evoked considerable discussion. A clinical diagnosis is rarely made, and even when recognized during life it is doubtful whether therapeutic measures have any distinct influence in arresting its progress. Huguenin's article in von Ziemssen's *Cyclopedia* still remains the classical monograph upon the subject, and during the past thirty years little of importance has been added.

The condition is rarely seen in the postmortem room of a general hospital, but is not uncommon in asylum practice. Thus in 1185 postmortems at the Government Hospital for the Insane, Washington, there were 197 cases with "a true neomembrane of internal pachymeningitis" (Blackburn). The disease is an affection of later life. Of the cases collected by Huguenin, 41 per cent. were between sixty and eighty, and 31 per cent. between forty and sixty years of age. During the first year of life 2.7 per cent. occurred; injury to the cranium during delivery no doubt accounts for this. Males suffer more frequently than females in the proportion of three to one (Durand-Fardel).

A great variety of associated morbid conditions have been met with. Among Blackburn's 197 cases in 45 there was chronic dementia, in 37 general paralysis, in 30 senile dementia, in 28 chronic mania, in 28 chronic melancholia, in 22 chronic epileptic insanity, in 6 acute mania, and in 1 case imbecility. Krönig, who analyzed 135 cases examined post-



mortem at the Berlin Pathological Institute, found phthisis (23 per cent.), general paralysis of the insane (19 per cent.), cardiac valvular disease (18 per cent.), syphilis (11 per cent.), puerperal sepsis (9 per cent.), and chronic alcoholism, carcinoma, and infectious diseases (about 6 per cent. each). Hemophilia and scurvy are also recognized causes, as are diseases of the blood, especially pernicious anemia. Traumatism is sometimes a determining factor (Schneider, 17 out of 74 cases). According to Huguenin, the great majority of cases occur with changes which cause a reduction in the volume of the brain, especially general paralysis of the insane. It is reasonable to suppose that the loss of support to the dural vessels which must accompany wasting of the brain predisposes to hemorrhage. Further, disease of the vessels is usually demonstrable in association with the pathological states which give rise to shrinkage of the cerebrum.

On postmortem examination there may be only a thin red or grayish-red film on the inner surface of the dura mater. The convexity of the cerebrum is its most common situation, although it may extend over a considerable extent of one or both hemispheres. Among 54 cases personally examined by Wigglesworth, 20 were strictly unilateral and 34 were bilateral, while 56 per cent. of the cases observed by Huguenin had a bilateral distribution. The membrane may extend to the base of the brain and is not uncommonly met with on the upper surface of the tentorium, although very rarely beneath it (Wigglesworth). In more advanced cases it may be of considerable thickness, consisting of a series of laminae which have been successively deposited, the most recent layers being next the cerebrum. There may be as many as twenty of these laminae, of which those of later origin are of a reddish color, the older layers being paler. Between the individual layers collections of fluid blood may be found. In some cases the false membrane may actually measure 2 or 3 cm. in thickness.

**Symptoms.**—These are various, and do not form a precise clinical picture. In many cases no symptoms are produced, and it may be that those which are present are obscured by those dependent on the original disease. A cerebral complication may be diagnosed, but its exact nature often remains a matter of uncertainty. The symptoms depend on the situation and extent of the morbid process. A degree of excitability with general restlessness, elevation of temperature, headache, giddiness, and vomiting may signalize the onset or extension of the lesion. The headache is sometimes very severe, and the patient may become delirious. Localized and recurring convulsive attacks, monoplegic or hemiplegic in distribution with, it may be, conjugate deviation of the eyes, paralysis or paresis of the hemiplegic or monoplegic type, slowing and irregularity of the pulse, and the gradual development of stupor and coma is a common history. The cranial nerves are rarely involved. Aphasia is occasionally present. There may be optic neuritis. Recovery may take place, to be followed by another attack.

**Diagnosis.**—The diagnosis may present the greatest difficulty. The condition may be suspected when in an individual who is suffering from one of the predisposing diseases, cerebral irritative phenomena develop

with associated paralysis, these symptoms varying from time to time and being accompanied by more or less stupefaction and, it may be, coma. Hemorrhagic fluid may be obtained on lumbar puncture in traumatic cases.

**Treatment.**—The head may be elevated and ice applied, but it is very doubtful whether any known treatment tends to arrest the bleeding.

**Pachymeningitis Cervicalis Hypertrophica (Hypertrophic Internal Pachymeningitis).**—This is characterized anatomically by great thickening of the spinal membranes, more especially in the cervical region, with consequent pressure upon the spinal cord and its nerve roots. In a typical case the lower cervical portion of the cord is found to be surrounded by a dense fibrous covering, which may be five or ten times (Oppenheim) as thick as the normal dura. The cord and its nerve roots are in fact embedded in an annular fusiform tumor of dense fibrous tissue of, it may be, the consistence of cartilage, usually thickest posteriorly and consisting of concentric layers inseparable from one another. The anatomical features are similar to those in chronic syphilitic meningo-myelitis. According to Wieter, the process actually originates in the leptomeninges, and should be named meningo-myelitis cervicalis chronica rather than pachymeningitis, since the dura is only secondarily involved. Pressure on the nerve roots induces necrotic changes in them with increase of interstitial tissue. The cord is flattened anteroposteriorly and ultimately ascending and descending degenerations are produced. Cavity formation, a consequence of intramedullary softening, is sometimes observed. It must not be supposed, however, that the process is limited to the cervical region. True, it is usually most pronounced here, but it often extends through a considerable extent and occasionally implicates the medulla and pons

There is little known with regard to *etiology*. Cold, injury, and over-exertion are all cited as possible causes. Syphilis appears to be an undoubted factor in some cases.

Neuralgic pains, often very severe, and referred to the distribution of the posterior roots entering the cord at the level of the lesion, are usually the earliest *symptoms*. This is the neuralgic stage of Charcot. At a later date, local and increasing muscular weakness, with wasting and impairment or loss of sensation, a consequence of interference with conduction in the spinal nerve roots, result. The small muscles of the hand and the flexors of the fingers and wrist are usually, because of the situation of the lesion, first affected, hence the deformity known as the "preacher's hand," which is not uncommon. Later still a third stage is reached, which is characterized by interference with conduction in the spinal cord, spastic paraplegia with increased reflexes, contractures, anesthesia of the lower extremities, loss of power over the bladder and rectum, and bedsores. When the process involves the medulla and pons, various cranial nerves may be implicated.

The very gradual development of severe bilateral pains in the neck and arms, succeeded by weakness, muscular wasting, and, it may be, objective sensory disturbance having a root distribution, and at a later date by symptoms of progressive pressure on the spinal cord, is a very

suggestive history when no evidence of vertebral disease can be detected.

Tuberculous pachymeningitis may present an identical clinical picture when there is no evidence of disease of the bone. The detection of tuberculous lesions in other parts of the body, an *x*-ray photograph of the spine, and the tuberculin test may be of value under these circumstances. When pressure symptoms are due to a simple tumor of the meninges, they are often unilateral.

Syringomyelia and amyotrophic lateral sclerosis may closely simulate hypertrophic pachymeningitis, but these conditions are unaccompanied by the severe pains in the neck and arms, which are so characteristic of the affection under consideration. The co-existence of syringomyelia and pachymeningitis has been noted.

A thorough antisyphilitic course of *treatment* is called for in any case in which there is a possibility of syphilis. Counterirritation with the cautery over the lower cervical vertebræ is sometimes useful. The various analgesics may be used to relieve the pains, which will often, however, only yield to morphine.

### ACUTE CEREBROSPINAL MENINGITIS.

**Etiology.**—Cerebrospinal fever, tuberculous meningitis, the acute affection of the meninges met with as an occasional sequel of syphilis, and the amicrobic serous meningitis of Quinke are described elsewhere. A group of cases, sometimes included under the comprehensive title of Acute Purulent Meningitis, remains for consideration. These may be subdivided according to their mode of production and with special reference to the causative agents upon which they depend. The causes are very varied, and yet the distribution of the inflammatory process, its character, and the accompanying symptoms present no constant features distinctive of the individual infections.

Leptomeningitis may be produced by many organisms, yet if we except the types due to the meningococcus, pneumococcus, streptococcus, and the tubercle bacillus, the general statement holds good that acute cerebrospinal meningitis is of rare occurrence. Councilman in 60 consecutive cases of the disease found the following organisms: Pneumococcus (18), streptococcus (18), meningococcus (21), staphylococcus (2).

Acute leptomeningitis may be of primary or secondary origin. The latter group of cases may be subdivided according to the conditions with which they are associated.

*Primary* leptomeningitis, apart from cerebrospinal fever, is of very rare occurrence, and is almost always due to the pneumococcus. Thus, Marchal, quoted by Weichselbaum, found that the meningococcus was responsible for 50.5 per cent. and the pneumococcus for 42 per cent. of the primary cases of sporadic meningitis. The mode of entrance of the organisms in primary meningitis is a debatable question. Infection by way of the nose is very probable in some cases. It is to be remembered that two factors are necessary in order that a primary inflammation of the meninges may be produced: (1) the infective organism must be pres-

ent, while (2) predisposing causes resulting in lowered resistance must also exist (Weichselbaum).

*Secondary* meningitis may be associated with a variety of conditions. Trauma is a not uncommon factor; thus a leptomeningitis may be directly produced by a penetrating wound or a compound fracture of the skull. The manner in which the organisms obtain access to the meninges under these circumstances is obvious. Infection through some of the accessory sinuses may account for cases in which a meningitis follows a fracture of the skull in which the skin surface has not been broken. Extension from adjacent disease is responsible for many cases. An acute or chronic otitis is much the most common factor in this connection. The pus may pass by direct extension through the bone, or the infection may spread by way of the sheaths of the facial or auditory nerves, or be carried by a suppurative phlebitis. When the infective process stops short of the dura an extradural abscess or a serous meningitis may result. If it perforates the dura an encapsuled meningeal or brain abscess or a general purulent meningitis follows. Exceptionally meningitis is secondary to disease in the nasopharynx, orbit, or frontal sinuses. Erysipelas of the scalp or a carbuncle in this situation are occasional causes. A generalized meningitis may be set up by an abscess of the brain, even though rupture has not taken place.

In the course of the *infective fevers* meningitis is met with as an occasional complication. It is to be remembered that a meningitis so occurring is not necessarily due to the same cause as the original disease, for mixed infections are not uncommon (Fürbringer). Pneumonia is the acute fever in which meningitis is most often met with. Osler found meningitis in 8 of 100 autopsies, while among 253 cases of pneumonia examined after death, Aufrecht met with 7 in which meningitis was present. Musser and Norris found a postmortem record of meningitis in 180 of 4833 cases of pneumonia. An acute meningitis is sometimes a terminal process in a general streptococcal infection. Its occurrence in erysipelas is much rarer than was at one time supposed. Thus Anders in his analysis of 1674 cases of this disease does not refer further to it than to remark that it is an occasional complication. Typhoid fever is very rarely complicated by true meningitis. Cole has collected 21 cases of meningism, serous and purulent meningitis, and 6 instances were present in the Johns Hopkins Hospital series of 1500 cases analyzed by McCrae. A few cases have been reported in which the meninges were affected without any other lesion. Dubois, in 1903, collected 11 cases in which a purulent meningitis was due to Pfeiffer's bacillus. The rarity of influenzal meningitis may be gathered from the fact that of 55,263 cases of this disease occurring in the German army meningitis was only noted in four. There are eight cases of primary suppurative meningitis in scarlet fever recorded in the literature (Teissier, Boudon and Duvoir, 1908). G. Henderson and W. T. Ritchie (1908) reviewed the reported cases of gonorrhoeal meningitis. Vennet (1908) collected the cases of meningitis occurring in association with mumps.

The occurrence of meningitis in measles, smallpox, ulcerative endocarditis, rheumatic fever, actinomycosis, and anthrax has also been recorded.

A streptothrix meningitis has been described. Lastly, acute meningitis may be associated as a terminal infection with such diseases as chronic nephritis, arteriosclerosis, heart disease, gout, and the wasting diseases of children (Osler).

**Pathology.**—Acute leptomeningitis is an inflammatory process which commonly implicates both the cerebral and spinal meninges. The degree and distribution vary; in some the base of the brain, in others the vertex is chiefly affected. The simple non-tuberculous meningitis of children is basal. Among Nauwerck's 27 pneumococcal cases the base alone was involved in 4, the convexity and base in 16, while in 7 the convexity, base, and cord were all implicated. When a leptomeningitis is secondary to adjacent disease, as, for example, otitis media, the process may remain for a time confined to the corresponding hemisphere, but in the great majority of cases, whatever the cause, the infection soon becomes general.

The earliest visible change is a hyperemia of the pia arachnoid, which is soon followed by some cloudiness of these membranes. Accompanying this is an excessive secretion of cerebrospinal fluid at first clear, later turbid, and ultimately, it may be, purulent. Inflammatory exudation occurs along the course of the engorged vessels. The yellow lymph may gradually spread from the sulci over the convolutions. Finally, the surface of the brain may be covered, as is not uncommon in pneumococcal cases, with a thick layer of greenish pus. It must not be supposed that the underlying brain escapes. An inflammatory cedema of the cortex is almost always present, and may go on to a true encephalitis with hemorrhages, hence the term meningo-encephalitis so often applied. Sooner or later a serous or seropurulent exudation occurs into the ventricles, the ependyma of which may be covered with lymph. When communication between the ventricles and subarachnoid space is interfered with, a degree of ventricular distension develops, which, however, rarely reaches the marked hydrocephalus met with in the tuberculous and serous variety.

The microscopic changes are practically identical with the appearances described under cerebrospinal fever. Engorgement of the meningeal and cortical vessels, the walls of which are infiltrated with leukocytes, is the most striking feature. The inflammatory process extends along the course of the bloodvessels into the brain substance. The neuroglia commonly shows some increase, while very pronounced changes in the pyramidal cells of the cortex are commonly present (Voisin). An acute proliferative inflammation of the veins and arteries is often seen in the forms of meningitis due to the pneumococcus and streptococcus (Councilman).

**Symptoms.**—The symptoms in acute purulent meningitis vary with the distribution, nature, and intensity of the process. Sometimes there is a premonitory stage in which such symptoms as irritability, restlessness, somnolence, malaise, headache, and vomiting are complained of. When the meningitis occurs in the course of an acute disease, such as pneumonia, the earlier symptoms, if slight, may be obscured by those of the primary affection. Sometimes the first indication is intense headache, cerebral vomiting, or a general convulsion, followed, it may

be, by delirium and rapidly deepening coma. A rigor is not uncommon at the outset, especially when the ventricular meninges are attacked.

*Headache*, frequently the earliest as well as the most distressing symptom, is rarely absent. It is, as a rule, widespread, although often referred especially to the frontal or occipital regions. When the meningitis is due to extension from adjacent disease, the headache may remain localized for a time. The pain is often very severe, the patient, even when in a semicomatose state, putting his hand to his head and crying out. "Cerebral vomiting," a common symptom, often develops at the outset and may persist. Occasionally it is absent. Delirium is often present and may appear at an early date, especially in vertical cases; it is sometimes violent. The patient is in some cases, even from the first, in a somnolent state, which gradually passes into coma. Photophobia and hypersensitiveness are common early symptoms, while cutaneous and muscular hyperesthesia is almost invariably present at some stage.

*Rigidity* of the neck, with pain on movement and perhaps head retraction, the latter symptom being rarely, however, so extreme as the retraction met with in the meningococcal form, is the rule in basal meningitis. When the meninges of the cord are prominently involved, rigidity of the back with opisthotonos may be observed.

The *pulse*, although increased in frequency, is usually slow at the commencement of the illness and not uncommonly irregular both in time and force. Later it is often small and rapid. Considerable variations in the pulse rate within a short period are often to be noted. A pulse rate which is unduly slow in comparison with the height of the temperature is a striking feature in some cases. The frequency of the respirations may be increased, and they are often irregular. Cheyne-Stokes breathing may be observed, especially in the event of an approaching fatal termination.

*Convulsions* of the Jacksonian type are not uncommon, while fits, unilateral or generalized, are prone to occur in children, and especially, according to most authors, in cases in which the meningitis is vertical in distribution. It is interesting to note in this connection that convulsions were not present in a single instance of the cases of pneumonia, ulcerative endocarditis, or septicemia complicated with meningitis observed by Osler. Choreic movements are very occasionally seen. Paralysis is far from infrequent in the later stages, and would doubtless be more often recognized were it not for the coma which often obscures it. Reinhold in 53 cases found hemiparesis fourteen times and a monoplegia on three occasions. The motor weakness may be associated with rigidity. Aphasia is an occasional symptom.

*Kernig's sign* is, as a rule, present; thus, Netter found it in 41 of 46 cases. This sign is by no means pathognomonic of a meningeal affection.

*Optic neuritis* is not infrequent in basal cases, although pronounced papillitis, such as is seen in cases of intracranial newgrowth, is very rare. The pupils are often unequal, and are apt to vary much in size from time to time. In the early stages they are usually contracted, while at a later date dilatation is the rule. Reflex iridoplegia is sometimes observed. Strabismus and ptosis are frequent, nystagmus not uncommon. Con-

jugate deviation of the eyes is occasionally met with. Paresis of the face, often fugitive, and accompanied at times by tremor in the affected muscles, is sometimes noted. Trismus and trophic ulceration of the cornea indicating involvement of the fifth nerve are more unusual symptoms. Dysphagia is by no means rare.

The tendon jerks, generally increased in the early stages, are usually lost later in the course. The plantar reflex may be of the extensor type. Evidence of vasomotor irritability is forthcoming in flushing of the skin and in the *tache cérébrale*, which is usually pronounced. Constipation is almost invariable. Sometimes there is retention of urine, while later there may be incontinence. Glycosuria has been recorded. Rapid emaciation is very often observed. The leukocyte count is commonly high although notable exceptions occur.

Upon *lumbar puncture* a turbid fluid, which emerges under pressure, is usually obtained, even in the early stages; occasionally, however, the fluid is quite clear, very rarely it is so thick it will not flow through the needle. The fluid is found to contain large numbers of cells, the great majority of which are polymorphonuclears, with, in addition, perhaps, the specific organisms which are responsible for the meningitis.

**Diagnosis.**—Acute leptomeningitis may be suspected when cerebral symptoms, such as headache, “cerebral vomiting,” irritability, general hyperesthesia, delirium, and convulsions, develop in association with the phenomena attendant on a febrile process, especially when these symptoms occur in conjunction with one of those conditions which are known to be of special causative moment. All these symptoms may, however, occur in the course of some of the specific fevers, notably pneumonia, typhoid, and influenza, as a result of congestion of the meninges. Rigidity of the neck and Kernig’s sign increase the probability of meningitis, while the additional presence of local palsies of the limbs or cranial nerves affords strong corroborative evidence.

Schultze holds that headache, rigidity of the neck, and hyperesthesia constitute a symptomatic triad, in the absence of which the diagnosis of meningitis is uncertain. Corroborative evidence is forthcoming if on lumbar puncture a turbid cerebrospinal fluid containing organisms is obtained. The various forms of acute generalized meningitis cannot be said to present symptomatic features, but the nature of a meningitis may be suspected when it occurs as a complication in the course of co-existing disease, and its pathology determined if specific organisms are detected in the cerebrospinal fluid.

*Meningism* or *pseudo-meningitis* is a term applied to a symptom complex met with in the course of some of the infective fevers, which is characterized by symptoms indistinguishable from the general symptoms of meningitis, and yet when opportunity is afforded of examining these cases postmortem no inflammatory changes are detected. In some of these cases there may be a slight degree of meningitis actually present which is recovered from; in many there is probably merely a meningeal hyperemia; while in others the symptoms would appear to be due to the direct effects of a toxin upon the brain itself. Meningism

may be distinguished from true meningitis by the fact that although the cerebrospinal fluid may emerge under pressure, it presents normal appearances.

*Hysteria* may simulate meningitis closely, and the diagnosis may be very difficult when hysterical symptoms occur in the course of an infective disorder. Tuberculous meningitis is, according to Gowers, especially apt to be mistaken for hysteria, while the reverse mistake is seldom made. Such signs of organic disease as optic neuritis, ankle clonus, Babinski's sign, and nystagmus are of great importance, while a turbid fluid obtained on lumbar puncture will determine the point.

*Acute suppurative otitis* sometimes presents a picture closely resembling that of acute meningitis. This is especially so in children, for delirium and convulsions, symptoms so apt to occur in early life in any acute febrile disturbance, may be accompanied by headache, vomiting, and giddiness due to the otitis. Here, again, lumbar puncture will be found to be of value.

An intracranial *abscess* may give rise to symptoms very similar to those of meningitis. Further, abscess and meningitis may co-exist. An acute onset, with high temperature, rapid pulse, hyperesthesia, irritative phenomena, and a high leukocytosis are in favor of the latter affection, as are head retraction, rigidity of the spine, and Kernig's sign. Symptoms pointing to focal disease in the cerebellum or temporo-sphenoidal lobe, *i. e.*, in the localities in which otitic abscess occurs, may indicate the presence of an abscess, but the absence of definite local manifestations does not exclude this possibility. When there is no co-existing ear disease the likelihood of abscess is small. In an uncomplicated case of abscess the cerebrospinal fluid is clear and contains few cells. The symptoms of infective sinus thrombosis may closely resemble those due to an otitic meningitis. A pyemic temperature and the occurrence of rigors are very suggestive of the first-named complication.

*Tuberculous meningitis* sometimes gives rise to difficulty, and a discharge from the ear does not exclude tuberculous meningitis, for the ear disease is sometimes due to the tubercle bacillus. Tuberculous lesions are usually present elsewhere in the body, hyperesthesia is commonly absent, and these cases, as a rule, run a more chronic course. The absence of a leukocytosis favors tubercle (Cabot found a leukocytosis in 32 of 43 cases of tuberculous meningitis). Choroidal tubercles when present make the diagnosis certain (Koplik found choroidal tubercles in 9 of the 46 cases in the first two days of the illness). The cerebrospinal fluid is often clear in tuberculous meningitis, and in contrast to the purulent form the increase of cells which is often present is found to consist almost entirely of mononuclear elements. Bernstein found the tubercle bacillus in the fluid obtained on lumbar puncture in 100 of 102 cases in which he looked for it.

The differential diagnosis of meningococcal meningitis from other forms of acute meningitis has been considered in a previous volume.

The delirium of alcohol and the uremic state may resemble acute meningitis, although it very rarely happens that the diagnostic problem is attended with much difficulty.



**Prognosis.**—The prospect of recovery from an acute purulent leptomeningitis is small. Death is almost invariable in cases with a sudden onset. In any form if the stage of coma has been reached, death is all but certain (Gowers). If the meningitis is localized the outlook is more favorable than when a general meningitis exists. The prognosis varies with the nature of the bacterial infection. The probability of recovery is greater in cerebrospinal fever than in purulent meningitis due to other causes. Recovery from pneumococcal meningitis, whether primary or secondary in origin, is most unusual. Thus of the 33 cases collected by Musser and Norris, 31 (93.3 per cent.) died, while only 1 of the 11 cases described by Cantley recovered. Death occurred in all of Nauwerck's cases. The chances of recovery are greater in a staphylococcal than in a streptococcal infection. The duration of acute meningitis varies from a few hours to two or three weeks. Of the 65 cases collected by Netter, 54 died within four days.

Macewen twenty years ago reported 12 cases of local otitic meningitis upon which he had operated, in 6 of which recovery took place. Cases of otitic meningitis, with pus and staphylococci in the cerebrospinal fluid, may recover after operative treatment of the purulent focus in the petrous bone (Körner, Alexander).

**Treatment.**—The management of cases of meningitis consists in the adoption of certain general measures and the alleviation of symptoms. The patient must be kept absolutely at rest in a quiet, darkened room; a dose of calomel may be given with possible advantage if the case is seen early. The head should be shaved and an ice-bag applied to it. Counterirritation is generally considered to be of service, and with this object a blister may be applied to the back of the neck. Hexamethylenamine may be given in doses of five to ten grains every four hours. There appears to be no evidence to show that injections of collargol and other substances into the subdural space are followed by any beneficial effect.

Headache and convulsions may be relieved by bromide and chloral. Morphine may be required when the headache is very severe. Vomiting is sometimes alleviated by a mustard leaf to the epigastrium or by giving the patient ice to suck. For general irritability, hyperesthesia, and delirium, warm baths are undoubtedly of value. When the temperature reaches an abnormally high point, tepid sponging should be employed.

When the meningitis is secondary to trauma or ear disease, surgical measures should be promptly resorted to.

Lumbar puncture is certainly of value in relieving the headache, and the patient may even be roused from a semicomatose state. The procedure is unattended with risk if care is taken not to lower the pressure of the cerebrospinal fluid below the normal. A marked amelioration of the symptoms may even follow the withdrawal of 10 cc.

**Chronic Leptomeningitis.**—The chronic inflammations of the meninges call for passing notice. Syphilis is the most common cause. A chronic inflammation of the meninges is also met with in confirmed alcoholics and in general paralysis of the insane. Tuberculous meningitis sometimes runs a chronic course. The duration in purulent meningitis is,

as we have seen, sometimes protracted, especially when the inflammation is localized, and necessarily in cases which ultimately recover. Posterior basic meningitis is a chronic disease, and in cerebrospinal fever the symptoms may persist for many months before death or recovery takes place. A localized chronic spinal leptomeningitis occurs in connection with caries of the spine and other forms of pressure paraplegia.

**Symptoms.**—These vary greatly and in the syphilitic and tuberculous forms often simulate closely those of tumor. The meningitis of chronic alcoholism may be suspected by its association rather than by any characteristic symptoms. The treatment varies with the cause.

**Localized Serous Spinal Meningitis.**—This is an affection which recently has attracted some attention. Horsley, who reviewed the literature of this affection under the name of chronic spinal meningitis, states that he has operated on 21 cases. A circumscribed collection of subarachnoid fluid is found pressing on the cord. In some cases it would appear that the condition is related to an inflammatory process, trauma, or syphilis; Bruns, however, is strongly of opinion that it may result as a primary idiopathic affection apart from disease of the spine, membranes, or cord. The *symptoms* are indistinguishable from those of tumor. Krause has collected 8 cases in which the Brown-Séquard syndrome and other symptoms led to laminectomy for the removal of a tumor, instead of which a localized serous meningitis was found. The same author, in 22 laminectomies performed for the removal of a supposed tumor, found in no less than six that a localized serous meningitis was the essential if not the only cause of the paralytic phenomena. In any case in which this condition is suspected an operation should be undertaken, since a subsidence of the symptoms usually follows.

**Lumbar Puncture and the Examination of the Cerebrospinal Fluid.**—The operation is a very simple one, it causes little pain, and with due precautions is unattended with risk. The patient sits astride a chair, over the back of which he leans, or, better still, he lies on his side with his knees fully flexed upon his abdomen and his spine well curved. The fourth lumbar space is that usually selected for the puncture. The cord cannot be injured, since it terminates opposite the second lumbar vertebra. Further, there is more room between the vertebræ here than at a higher level, while another reason for choosing this situation is that the dura is more firmly adherent to the bone here than elsewhere, hence there is little chance of pushing the membrane in front of the needle. A line joining the highest point of the iliac crests passes between the fourth and fifth lumbar spines. The needle should be three and a half inches in length and made of steel or of platinum with an iridium point, the advantage of the latter being that it can be boiled. The skin should be very carefully sterilized. A general anesthetic is unnecessary except in the case of excitable patients, but ethyl chloride may be used locally.

The needle is introduced midway between the fourth and fifth lumbar spines a quarter of an inch to one side of the middle line, and is passed almost directly forward with a slight inclination inward and upward. When the needle reaches the intervertebral ligament it receives a momentary check, but gentle, firm pressure sends it through the ligament and

dura, and its point then lies in the subarachnoid reservoir. The distance traversed by the needle before it reaches the dura varies from 2 to 7 cm., according to the age and development of the patient. Should the needle come in contact with bone, the point should be withdrawn for a short distance and passed in at a rather different angle. If after several attempts it is found impossible to avoid the bone, it is best to withdraw the needle altogether and reintroduce it in another situation. It not uncommonly happens that no fluid comes, even though the needle has been felt to pass through the ligament. Under these circumstances suction is not to be used, but a stilette should be passed along the needle, the lumen of which has probably been blocked on its way through the skin and muscles. Occasionally after this manipulation no fluid is obtained. This may be accounted for by the fact that the dura has been pushed in front of the needle, which has failed to penetrate it, or it may be that, as in some cases of hydrocephalus, the ventricular system has been shut off and there is no fluid in the subarachnoid space. Again, in some cases of purulent meningitis the fluid is so thick that it will not flow. Should the needle come in contact with a nerve root as it passes through the ligament, the patient complains of intense pain often radiating down the leg. The needle should then be withdrawn and reinserted.

Unpleasant effects very seldom follow the operation, provided that only a small quantity of fluid is withdrawn. Headache, sickness and faintness are sometimes complained of, hence it is advisable that the patient should rest for an hour after the procedure. A rest of twenty-four hours after the operation, advocated by Nissl, appears in the writer's experience to be unnecessary. A number of fatal results have been recorded after lumbar puncture in cases of intracranial tumor in which there was a great increase of intracranial pressure. Almost all these fatalities took place before the importance of withdrawing only a small quantity of fluid was realized. Nevertheless, lumbar puncture should only be undertaken after very careful consideration in these cases.

Under normal circumstances the fluid emerges drop by drop and is collected in a sterilized test-tube. The first few drops are sometimes turbid, owing to the fact that a little blood has been collected in the lumen of the needle in its passage through the muscles and skin; it is therefore advisable to allow this to escape.

The *pressure* under which the fluid emerges is of some importance. When the pressure is raised, the fluid, instead of emerging in drops, may spurt out in a steady stream. Quinke states that the normal pressure equals on the average a column of water of 40 to 60 mm., but that it can only be regarded as pathological when it is above 150 mm. Krönig, on the other hand, finds the average normal pressure in the horizontal posture to be from 100 to 150 mm., while in the sitting position he puts the average at 410 mm. According to the latter writer the pressure may rise to 700 mm. in pathological states. The pressure is often very greatly raised in cases of intracranial tumor and in the various forms of meningitis. It may also be increased in uremia and in certain of the acute infective fevers. According to Quinke moderate increase of pressure with severe pressure symptoms points to an acute, while a great rise of pressure with slight pressure symptoms indicates a chronic process.

Valuable information is often derived from the naked-eye appearance of the fluid. Thus in meningeal hemorrhage, and in hemorrhage into the ventricles, the fluid may be blood-stained. If such fluid is obtained on lumbar puncture, the question arises, Is the color of the fluid to be explained by puncture of a bloodvessel during the passage of the needle? If the fluid is received in three different tubes, and it is found that the tint of the three is the same, probably the blood is not due to the puncture (Campbell). Again, when hemorrhage has occurred into the cerebrospinal fluid, after the fluid is centrifuged it often retains a yellowish color, whereas when the hemorrhage is accidental it presents its normal appearance (Mathieu). Hematoidin crystals may be found in cases in which the hemorrhage is of some standing (Krönig). The fluid may be purulent; indeed, in some cases thick pus is withdrawn. A purulent cerebrospinal fluid means a purulent meningitis or a ruptured abscess. Even when there is a purulent meningitis the fluid obtained by lumbar puncture may be free from leukocytes and organisms, no doubt because communication between the cerebral and spinal subarachnoid space has been interfered with. In abscess the fluid is clear unless a meningitis either localized or generalized co-exists. The presence of a turbid fluid does not insure a certain diagnosis between general meningitis and a localized inflammation of the meninges associated with brain abscess, sinus phlebitis, and purulent disease of the labyrinth, for in all of these cases leukocytes and bacteria may pass into the cerebrospinal fluid. The cerebrospinal fluid in tuberculous meningitis may be quite transparent, although it usually shows some turbidity. A yellow coloration of the fluid has been observed in jaundice.

The *chemical* examination yields some information of diagnostic importance. The normal fluid contains a trace of serum globulin, and when boiled becomes slightly cloudy. An abnormal excess of the albuminous content is met with in acute meningitis, tabes, general paralysis of the insane, and active cerebrospinal syphilis. The amount of albumin present may be estimated by special methods. Fibrin, although absent from the normal fluid, is present in acute meningitis. The reducing substance, which is constantly present in the normal fluid and in the examination for which at least 10 cc. should be used, is, according to Bernstein, always absent in purulent meningitis, while in the tuberculous variety, although present in small quantities at the commencement, it disappears in the later stages. Considerable quantities of sugar have been found in the cerebrospinal fluid in diabetes. Cholin, a product of myelin degeneration, has been detected in the blood and cerebrospinal fluid by Mott and Halliburton in acute degenerative diseases of the nervous system.

The *cytological* examination yields valuable data. The same technique is to be constantly employed for comparative purposes. That originally recommended by Widal, Sicard, and Ravault is as follows: Three cubic centimeters of the fluid are withdrawn; this is centrifuged for ten minutes in an instrument that revolves 3000 times per minute; the tube is inverted for two minutes, the bottom of the tube is then scraped with a fine capillary pipette (a vaccine tube does excellently for this purpose), the fluid is blown on to a slide, the deposit is allowed to dry in the air,

it is fixed in equal parts of alcohol and ether for twenty minutes, and stained with methylene blue, eosin, Jenner's or Ehrlich's stain. Following this procedure it is found that under a magnification of 400 diameters the normal fluid shows not more than half a dozen cells to the field. If the normal fluid is examined in a counting chamber it is found that it does not contain more than two cells to the cubic millimeter.

The observations of importance in the examination of the film are (1) the number of leukocytes, (2) the types of cells present and their relative proportions, and (3) the presence of other elements. Both in epidemic meningitis and in the group of cases included in the present article there is commonly a great excess of cellular elements, and the increase is found to be largely due to polymorphonuclear cells. In long-standing cases in which recovery is taking place the mononuclear cells are found to preponderate. Very exceptionally a fluid containing no excess of cells is met with in acute meningitis.

Tuberculous meningitis differs from the other forms of cerebrospinal inflammation in that although the cells in the fluid are increased the increase is almost entirely due to an increase in the mononuclear leukocytes, and may be described as a lymphocytosis. Cases of acute tuberculous meningitis are occasionally met with in which the polymorphonuclear cells are in excess, but these are of rare occurrence.

In general paralysis of the insane, in tabes dorsalis, and in cerebrospinal syphilis a pronounced cerebrospinal lymphocytosis is almost invariable, and is usually most marked in the earlier stages of the disease. Thus, Purves Stewart found in eleven cases of general paralysis of the insane an average count of 127.5 lymphocytes per field, while in 22 tabetics the number in each field averaged 131.4. Very occasionally a case of tabes is met with in which there is no lymphocytosis. The presence of a lymphocytosis in an individual who has had syphilis points to implication of the central nervous system, and is an indication for treatment. A pronounced diminution in the number of cells may occur during antisyphilitic treatment. In cases of syphilis in which the nervous system is not affected there is no cerebrospinal lymphocytosis. A lymphocytosis has also been found to occur in cases of herpes zoster, very occasionally in cases of intracranial tumor, in chronic alcoholism, and in mumps. When present it is to be regarded as an expression of chronic meningeal irritation. Tumor cells have been detected in occasional cases of malignant tumor of the meninges.

The *bacteriological* examination is sometimes of great value. The detection of microorganisms enables us to exclude conditions in which bacteria are not present, and also to distinguish between the various forms of meningitis. In tuberculous meningitis the bacillus may be found in the large proportion of cases if only sufficient care is used. The pneumococcus, streptococcus, and meningococcus are the organisms most often found in meningitis. A number of instances of mixed infection have been reported. The Wassermann reaction in the cerebrospinal fluid has come to be regarded as of great importance in the diagnosis of nervous syphilis, for a positive reaction may be obtained in this fluid when that in the blood is negative.

## CHAPTER VI.

### DIFFUSE AND FOCAL DISEASES OF THE SPINAL CORD.

By E. FARQUHAR BUZZARD, M.D.

#### 1. THE LOCALIZATION OF NON-SYSTEMIC DISEASE AFFECTING THE SPINAL CORD AND CAUDA EQUINA.

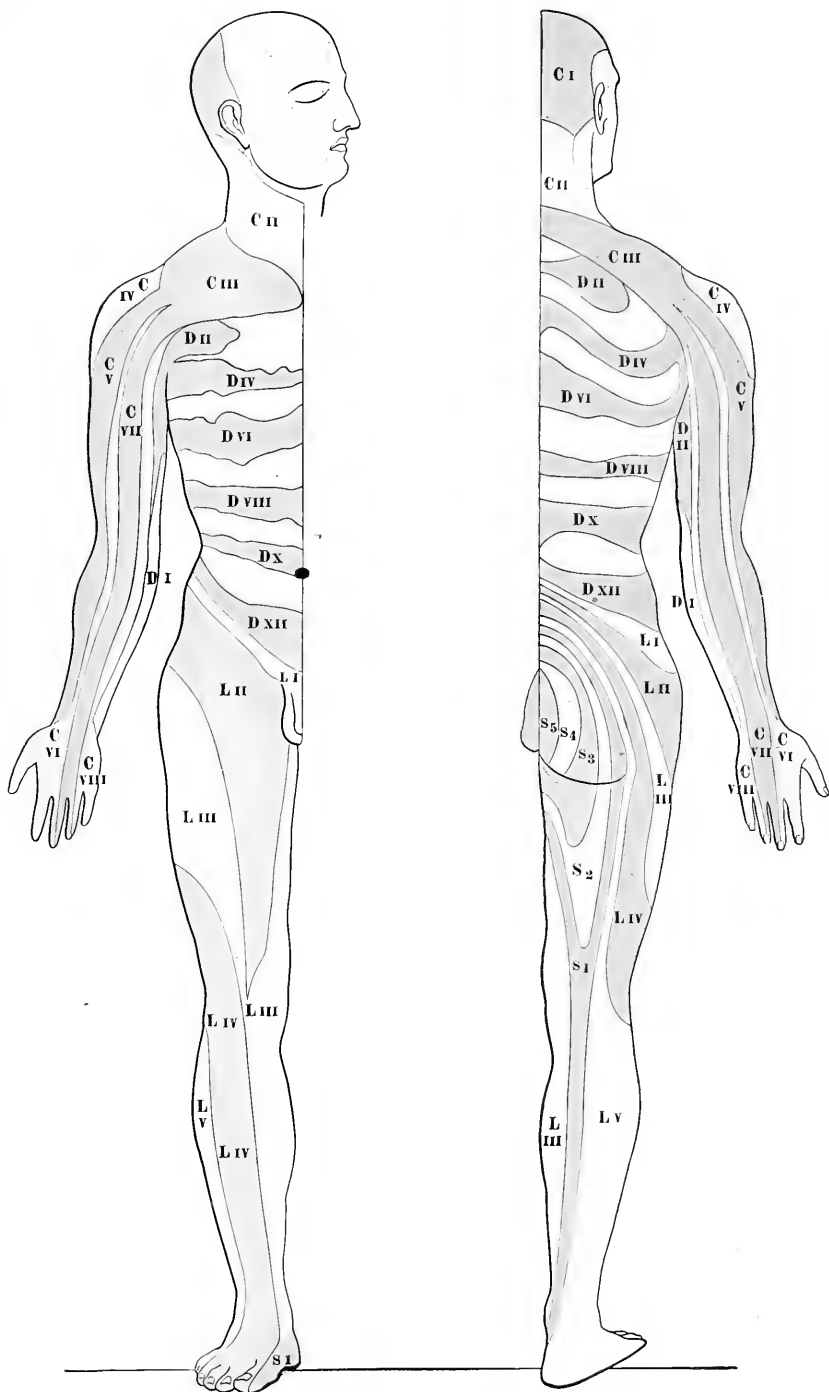
**Segmental Localization.**—In discussing the various morbid conditions which affect the spinal cord, it will be necessary to state on more than one occasion that the symptoms produced by a particular disease will depend upon the segmental level of the lesion. In order to avoid repetition, it is expedient to review the data upon which the diagnosis of the segmental level of disease is based. A *focal lesion* may be regarded as one which is limited longitudinally to one, two, or three segments of the cord, but which may involve either a part or the whole of the transverse area of those segments. A *diffuse lesion* may be defined as one which affects in a confluent or discrete fashion a considerable length of the cord, amounting to many segments. In neither case have we to deal in this section with morbid processes confined to one or more neuronie systems. From a clinical standpoint, when a diffuse lesion involves a considerable length of the spinal cord it is often difficult and sometimes impossible to do more than define the upper limit of the affected area. For this reason a focal lesion limited to a single segment is the most convenient proposition upon which to base a discussion of segmental localization.

Each segment of the spinal cord has two chief functions; in the first place, it forms a link in the chain of nervous paths which traverse it, the conduction of impulses along these paths being dependent upon the normal condition of the fibres within the limits of the segment; in the second place it is a centre for various activities of motor, sensory, vasomotor, trophic, or reflex character, all of which rely upon the integrity of the segment, and especially of the gray matter, together with the anterior and posterior roots, for their healthy expression.

**The Segment as a Link in the Chain of Nerve Tracts.**—The result of a lesion affecting the conducting paths depends upon the extent of interference with the function of these paths.

**Total Flaccid Paralysis.**—If the injury to a spinal segment is so severe that there is complete severance of all impulses, a condition of total flaccid paralysis supervenes in all parts innervated from the spinal cord below the level of the lesion. The motor palsy is associated with absolute loss of appreciation of all sensory impressions arising from the skin and other tissues supplied by the same region of the cord, and with abolition of all tendon reflexes in the same parts, at any rate for a

# PLATE IV



Areas of Anæsthesia upon the Body after Lesions in the Various Segments of the Spinal Cord.

The segments of the cord are numbered: C I to VIII, D I to XII, L I to V, S I to 5, and these numbers are placed on the region of the skin supplied by the sensory nerves of the corresponding segment.





considerable period of time. The loss of tendon reflexes is combined with absence of superficial reflexes, with the exception that an extensor response may in some cases be obtained on stimulating the soles of the feet. It is not quite certain whether such survival of the plantar reflex denotes or does not denote an incomplete physiological discontinuity of all impulses at the site of the lesion. Visceral reflexes are also interfered with, and complete incontinence of urine and feces rapidly supervenes, although this may be preceded, especially in young subjects, by a short period during which reflex evacuation takes place. There is cessation of all sexual power or desire, with a tendency to incomplete priapism in certain male patients. The skin of the paralyzed parts is liable to trophic changes, and if the lesion is acute there is often a period of vasodilatation with cedema, which is, however, soon replaced by vasoconstriction and a dry exfoliative cutis. In the early stages the muscles supplied from the cord below the lesion, although completely paralyzed and flaccid, preserve their nutrition and their normal reaction to electrical and mechanical stimuli. When the complete transverse lesion is of long duration an atrophic process gradually attacks the paralyzed muscles which ultimately lose their faradic excitability.

**Spastic Paralysis.**—If the interference is less profound, and yet sufficient materially to affect the conductivity of the chief motor and sensory tracts, the flaccid paralysis is replaced by a spastic paralysis of all muscles supplied by the cord below the level of the lesion. This spastic paralysis, which is sometimes known as “upper motor neuron paralysis,” and which we attribute to changes in the pyramidal tracts, is characterized by the presence of some degree of tonic muscular spasm or rigidity, often associated with a tendency to involuntary reflex spasms of a painful type. The muscles retain their nutrition, size, and normal electrical excitability, but their increased tone (hypertonicity) offers some resistance to passive movements. The amount of voluntary power varies with the severity of the lesion, but the relationship of degree of spasticity to degree of paresis is one the laws of which are not yet perfectly understood. The tendon reflexes of all the spastic muscles are increased and there is a tendency to the production of clonus. On the other hand, the superficial skin reflexes are diminished or abolished below the level of the lesion, with the exception that a plantar response of extensor type is usually present on stimulating the sole of the foot, and a cremasteric reflex is occasionally obtainable. With spastic paralysis the sensory loss may vary from complete insensibility to one in which only very fine tests are capable of eliciting some evidence of anesthesia in the skin supplied by segments below the level of the lesion. The intensity of anesthesia is often not equal all over this area, but further reference will be made to this later on. The condition of the sphincters is also variable, the slight cases generally showing, after an initial retention, some hesitancy or precipitancy of micturition, with constipation of the bowels; the severe cases, retention or incontinence of urine, associated with constipation and rectal incontinence after aperients. At the same time the sexual powers of the male are diminished or in abeyance.

When spastic paralysis of the lower limbs has been of long duration it

tends to the production of certain deformities, of which a pes cavus is the most common. Permanent contracture of muscles in the position of flexor spasm is another troublesome complication.

**The Segment as a Centre of Activity.**—The destruction of the gray matter by a focal lesion gives rise to certain phenomena distinct from those which result from the destruction of the conducting paths in the white matter. Each segment contains anterior horn cells, from which originate the anterior root fibres. As a result of the degeneration of these ganglion cells the efferent nerve roots degenerate, and the muscles innervated by them are paralyzed and rapidly waste. Fibrillary tremors or twitchings and a tendency to the formation of contractures are additional characteristics of what is known as “lower motor neuron paralysis.” If the muscles are examined electrically during this process of atrophy they are found to undergo the reaction of degeneration, *i. e.*, diminution and abolition of response to faradism, together with an altered slow response to galvanism. In this respect they differ from the paralyzed but spastic muscles supplied by nerves arising from segments below the focal lesion, which retain their size and nutrition and present no alteration in electrical excitability. The tendon reflexes of the muscle affected by the atrophic palsy disappear, as do all the reflexes subserved by the segment which is the seat of the disease. In the skin of the area innervated by the same segment, vasomotor and trophic changes are also to be noted, a moist, sodden surface, with atrophy of nails or other structures, being the usual result. Sensibility in the skin area from which the afferent fibres of the corresponding posterior root arise, is altered, but the anesthesia is less marked than the analgesia and is not so complete in the upper part of the root area as it is in the lower and in the areas subserved by the segments of the cord below the lesion. A zone of hyperesthesia may under certain conditions be present just above the anesthetic level. Should the segment involved by the focal lesion innervate the anal or vesical sphincter, the latter becomes toneless.

Speaking generally, the results of a severe transverse or focal lesion of the spinal cord are atrophic paralysis of the muscles supplied by the segment in which the lesion is situated, together with a spastic paralysis of the muscles innervated by segments below the level of the disease. Alteration of sensibility is present as high as the area supplied by the affected segment. In order to apply the above data to the localization of a spinal lesion, it is obviously necessary to know which muscles are supplied by each segment of the cord, the skin area from which sensory impulses reach each segment of the cord, and the situation of certain centres subserving visceral and other reflexes. The large majority of muscles receive fibres from two or more spinal segments, and a knowledge of their peripheral nerve supply is therefore but a poor indication of their spinal supply. There is the same discrepancy between the nervous and spinal supply in the segmental sensory areas, which are entirely different from the areas of distribution of peripheral nerves. In addition there are factors in segmental localization which deserve discussion.

**Spinal Root Symptoms.**—The recognition that certain symptoms may be dependent on a lesion of a spinal root rather than of a spinal segment

may be of immense clinical service, especially when the root concerned is a large one. It must be remembered that the result of an injury to the root may be much the same whether its site is close to the spinal origin of the root or at the point where it leaves the neural canal, a difference, it may be, of several inches. Root symptoms are sensory, motor, reflex, trophic, and vasomotor in character, but it is only by means of the sensory and trophic phenomena that any marked distinction can be drawn between a lesion of a spinal segment and one which involves its anterior and posterior roots.

In the first place, severe pain of a sharp, stabbing kind, referred to a segmental skin area, is suggestive of a root lesion, and this may be associated with some degree of hyperesthesia in the same region. In the second place, a destructive lesion of a single posterior spinal root may give rise to slight disturbances of sensibility of a certain character. For instance, the section of a first dorsal posterior root will produce some loss of painful and gross thermal sensibility over a region on the ulnar side of the forearm, which is smaller than the whole area supplied by the first dorsal root. The slighter degree of tactile anesthesia in the same area is due to the greater overlapping in contiguous roots of fibres subserving touch compared to those which are concerned with painful and gross thermal sensibility. From a localizing standpoint the presence of herpes zoster within a segmental skin area denotes disease involving the corresponding spinal root ganglion, and affords a valuable guide to the site of the lesion in certain cases in which the spinal column or meninges are affected. Sometimes a subcutaneous hemorrhage or other skin affection of similar distribution may appear as an indication of a root lesion. Motor and reflex activities are affected in a similar manner whether the anterior gray matter of the segment or its efferent roots are the seat of disease.

**Oculopupillary Symptoms.**—A number of fibres pass down from a centre in the brain-stem through the cervical cord in order to join the cervical sympathetic system, leaving the cord by the eighth cervical, the first and second dorsal nerve roots. In some lesions of the cervical cord, but more often in focal lesions of the first dorsal segment, injury to these fibres causes diminution in size of the palpebral fissure and of the pupil of the same side. In such instances of sympathetic miosis and ptosis the pupil may not actively dilate to shade or to the influence of cocaine. Irritation of the same system of fibres produces a reverse condition of mydriasis and exophthalmos.

**Optic Neuritis.**—In lesions of the cervical and upper dorsal cord the occurrence of a mild and occasionally of a severe degree of papillitis has been noted, but its mechanism has not been adequately explained.

**Vasomotor and Secretory Phenomena.**—An acute transverse lesion often produces temporary cutaneous vasodilatation and hyperidrosis in the paralyzed parts, and this may sometimes be associated with oedema of the skin, the deeper tissues, and occasionally of the synovial cavities. The fact that this is rapidly followed by complete secretory paralysis has been utilized by Horsley for the purpose of demarcating the upper limit of the affected skin by means of a dose of pilocarpine. A successful result is

sometimes obtained in cases of not very recent origin. Paralysis of the cervical sympathetic, with hyperemia and hyperidrosis of the face and neck, is met with in lesions of the cervical and upper dorsal segments.

**Cardiac and Respiratory Symptoms.**—Sympathetic fibres for the cardiac and pulmonary plexuses leave the upper four segments of the dorsal cord; lesions above or involving that region give rise to modification of cardiac and probably also of respiratory action, although the latter is difficult to estimate on account of the associated interference with the innervation of the respiratory musculature. It is important to remember that the diaphragm is innervated from the fourth cervical segment, and that lesions of that or higher segments affect the thoracic and abdominal respiratory muscles.

**Splanchnic Symptoms.**—The superior splanchnic nerve arises from fibres leaving the dorsal cord from the fifth to ninth segments. Injuries to these fibres in their intramedullary course is attended as a rule with marked but often only temporary tympanitic distension of the hollow abdominal viscera.

**Vesical, Rectal and Sexual Reflexes.**—The generally accepted view that there are definite centres in the lumbosacral enlargement of the cord connected with the vesical, rectal, and sexual reflexes has been strongly controverted recently on experimental and other grounds by Müller, who maintains that the central mechanism subserving these functions is situated in the sympathetic system. It would be out of place to discuss here the relative merits of these views, since the question is by no means settled; at the same time, the confession must be made that some of the clinical phenomena associated with the sphincters in spinal and cerebral disease have never received satisfactory explanation. Müller experimenting on dogs, found that after extirpation of the lumbosacral cord, periodic evacuations of the bladder and rectum were soon established after a short period of retention, and that in male animals erection of the penis, associated even with ejaculation, was still possible. The voluntary muscles of the external anal sphincter and of the higher parts of the urethra become toneless and flaccid when the lumbosacral cord is destroyed, but if the lesion only isolates this part of the cord from the higher centres the striated muscles retain their tone and also their reflex action, although it may be in a modified manner. According to Müller the lumbosacral cord is only necessary for the conduction of sensory impulses from the organs to the brain and for the conduction of voluntary impulses to the striated muscles just mentioned. Periodic contractions of the non-striated muscles of these parts are only dependent on the sympathetic system, just as normal emptying of the pregnant uterus will take place in complete motor and sensory paralysis of the lower part of the body. It is highly probable that the simplicity of the mechanism is complicated in human conditions by the unnatural supine position of the patient and the paralysis of other voluntary muscles which are normally concerned with the evacuation of the bladder and rectum, as well as by the common secondary changes in the vesical walls resulting from sepsis.

**Pain as a Localizing Symptom.**—This is a subject of much importance and difficulty in actual practice. It is probably true that focal or diffuse disease of the cord never gives rise to pain directly, but frequently does so indirectly by involvement of the osseous or meningeal coverings or posterior nerve roots. For this reason it is obvious that pain is more often associated with morbid processes which attack the cord from without than with those which originate in its own substance. It is necessary, however, to distinguish carefully between several forms of pain. A dull aching localized pain in the back is often present in cases when the spinal cord and theca are undergoing steady, slow compression, or, more rarely, when an intramedullary growth is causing a focal expansion of the cord of considerable size. This form of pain may be increased by movement of the vertebral column, and it may spontaneously undergo paroxysmal exacerbations of a burning character. It is generally referred to a point only slightly below the level of the lesion.

Sharp, shooting pains referred to particular regions, it may be at some distance from the spine, are characteristic of disease implicating the posterior spinal roots, and their accurate study may be of considerable value in determining the seat of injury. These pains may also be excited or intensified by movements, especially when the disease actually involves the vertebræ. In considering these root pains two important facts must be borne in mind; in the first place, the region to which the pain is referred will correspond to the cutaneous area supplied by that root; in the second place, the area of referred pain will be the same whether the root is affected near its medullary origin or in its course through the intervertebral foramen, this being of great significance when the root is one of considerable length. In the latter case, associated symptoms will aid in localizing the injury to the root more exactly. Care must be exercised in investigating the character of so-called girdle pain, and also of the pain which is associated with flexor spasms. Girdle pain is in some cases a root pain distributed through one of the belt-like dorsal segmental areas; in others it is a sense of constriction, which is often marked at the upper level of spasticity and anesthesia. Pain and cramps, associated with flexor spasms of the hips and knees, are generally referred to the front of the hip and back of the knee, and must not be confounded with the pain originating from injury to the lumbrosacral roots.

**Disturbances of Sensibility.**—Hitherto the disturbances of sensibility in transverse lesions of the spinal cord have been merely referred to as complete or incomplete anesthesia to various stimuli, extending over all parts of the body innervated from the cord below the site of disease. This may be sufficient when the lesion is really transverse, and influences equally all afferent paths as they traverse the affected segment, but it is totally inadequate to describe the various forms of sensory loss which may arise in connection with less complete and more patchy focal lesions, and especially those which are associated either with disease of the central gray matter or with unilateral morbid processes. For the proper understanding of such conditions reference must be made to the work of Head and Thompson on the grouping of afferent impulses within the spinal cord.

Head, in conjunction with Rivers, had previously shown that the

afferent mechanism in the peripheral nervous system consists of three systems:

1. *Deep Sensibility*.—This conveys impulses excited by pressure and by all movements of joints, tendons, and muscles. Painful impulses derived from excessive pressure are also carried by this system. The patient in whom this system is intact is not only able to recognize movements of joints, but also the locality of the stimulus and the direction of the movement. The fibres which conduct these sensory impulses run mainly with the muscular nerves, and are not destroyed by division of all the sensory nerves to the skin.

2. *Protopathic Sensibility*.—This system is capable of responding to painful cutaneous stimuli and to the more extreme degrees of heat and cold. The appreciation of these stimuli is diffuse and inexact as to the locality of the spot stimulated.

3. *Epicritic Sensibility*.—To this system is due the power of perceiving and locating light touches, of discriminating between two points applied simultaneously to the surface, and of recognizing the finer grades of temperature, called cool and warm.

Head and Thompson have shown that the impulses of these three different systems combine in new groups very soon after they enter the spinal cord. Some impulses cross to the opposite side immediately, others cross after running a short course on the same side, and others ascend to the upper extremity of the cord entirely on the side of their entry. This re-arrangement of afferent impulses and their rough position in their spinal paths may be briefly summarized in the following manner:

1. Impulses of pain, whether excited by cutaneous stimuli or by excessive pressure, run together in the spinal cord and cross, probably early, to the opposite side.

2. Impulses of temperature of *all degrees* are combined, cross to the opposite side, and are closely associated, but not intermingled, with those of pain. Impulses of heat are also separated from those of cold.

3. Impulses excited by light touch and by pressure and those which subserve their localization accompany each other, cross to the opposite side, probably less rapidly and less completely than those of pain and temperature, and ascend in a path which is distinct from that of the latter.

4. Impulses subserving the sense of passive position and movement are associated with those of tactile discrimination (compass points) in their ascent of the cord on the same side as their entry. These impulses probably pass up the posterior columns.

5. Of the non-sensory afferent impulses, many ascend in the secondary system of the direct cerebellar tract to reach the cerebellum.

Head and Thompson then state their belief that the recombination of afferent impulses into new groups takes place on the side by which they enter the spinal cord, and that all those impulses which cross to the opposite side are carried by systems of fibres belonging to a secondary or intramedullary level. On the other hand, the impulses subserving the sense of passive position and movement and of tactile discrimination remain in a system belonging to the primary or peripheral level in their ascent on the same side of the cord.

In the light of their observations, the symptom complex which is known by the name of Brown-Séquard's paralysis may now be considered. He observed that a hemisection of the spinal cord was followed by motor paralysis and loss of muscular sense on the side of the lesion, and by loss of other forms of sensibility at the level of the lesion on the same side, and below the level of the lesion on the opposite side of the body. Well-defined local lesions of one-half of the spinal cord are of rare occurrence, but modified forms of Brown-Séquard's paralysis are not infrequent and those which have been examined bear out in the main the conclusions

FIG. 13

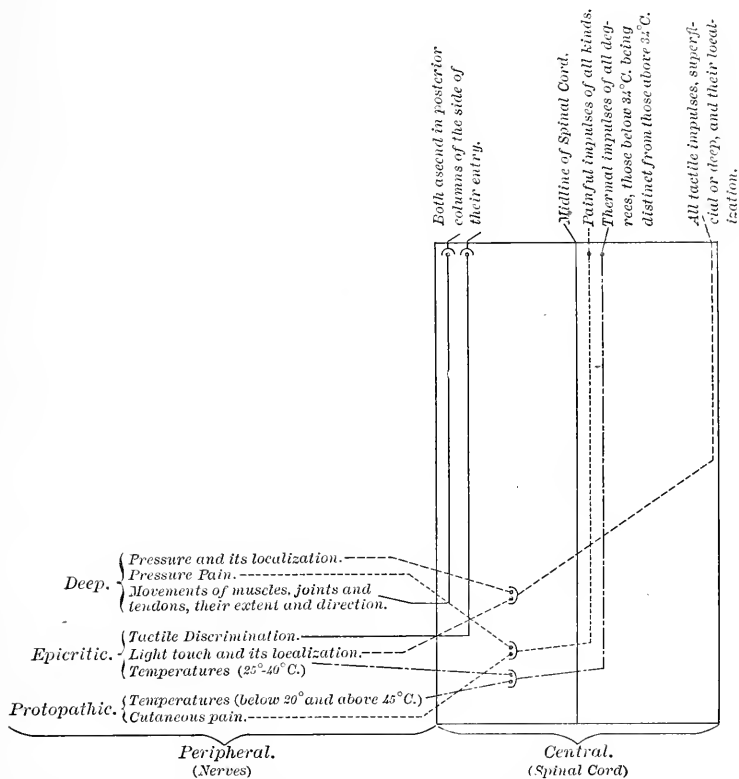


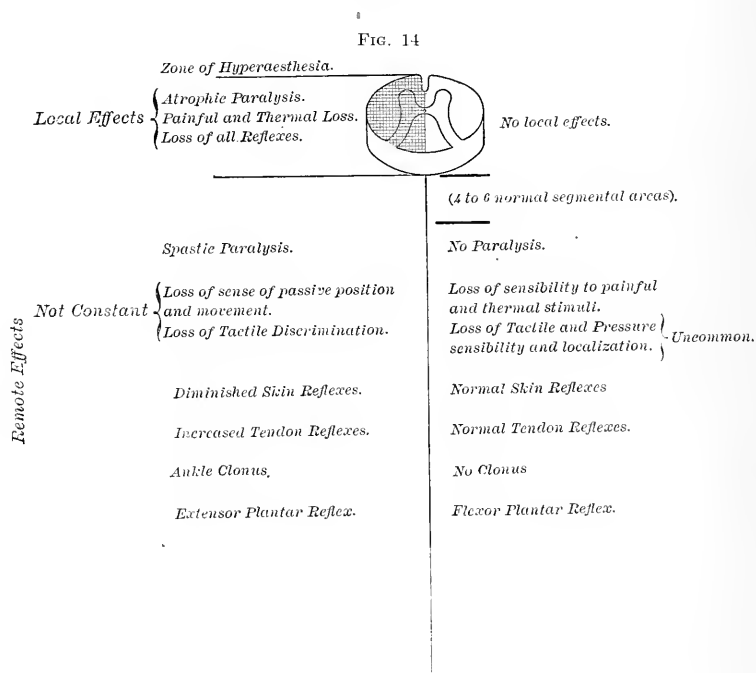
Diagram illustrating the grouping of afferent impulses within the spinal cord according to Head and Thompson.

of Brown-Séquard. With improved methods of examination in relation not only to sensory, but to motor and reflex disturbances, a perfect hemisection of the cord should produce a condition which may be graphically represented in the diagram (Fig. 14).

If an exception be made for those phenomena marked "uncommon" or "not constant" in the diagram, the remaining symptoms present a picture which is by no means rare in focal diseases of the spinal cord.

Apart from the sensory disturbances connected with unilateral lesions of the cord, there are one or two other points of interest. It is by no

means rare to find that the loss of cutaneous sensibility below an incomplete transverse lesion of the cord is not equal in all the skin areas which are affected. In the first place it may happen that when pressure is slowly applied to the spinal cord at the level, for instance, of the second dorsal segment, the sensory loss in the early stages may only be traced as high as the mid-dorsal segmental areas, or may only be complete up to that level and relative in those above that level. This is readily understood when it is remembered that the long secondary afferent paths lying in the periphery of the cord are first affected by pressure, and that the decussation of sensory impulses is a gradual process not completed in less than four, five, or six segments.



In the second place, when the upper limit of sensory loss is clear and well defined, and corresponds to the level of the lesion, it may be found that some of the lower sacral areas have escaped the anesthesia of surrounding regions. Such a phenomenon can only be explained, as Head points out, by a lamellar arrangement of the segmental impulses as they pass up the spinal cord.

**Diagnosis of Disease of the Cauda Equina.**—The cauda equina is contained within the vertebral canal from the second lumbar vertebra to the coccyx, losing a pair of roots at every intervertebral space on either side. In certain cases there arises a difficulty in distinguishing between lesions of the lumbosacral segments and those of the cauda equina. Certain principles must be remembered in this connection. In both segmental and root lesions the symptoms may be similar, the distribution of motor and sensory loss being of the segmental type. A lesion of a



lumbar segment, however, produces symptoms relating not only to that segment, but to each segment below it, while a lesion of a lumbar root may be distinct from any involvement of the roots or segments above or below.

It is clear that all the sensory and motor symptoms are of the spinal root type in lesions of the cauda equina, and that the presence of any spasticity, any increase in the deep reflexes, clonus, or an extensor plantar response, negatives the diagnosis of disease limited to that region. All muscles which are paralyzed are also wasted, have undergone the reaction of degeneration, and have lost their tendon reflexes. When the sphincters are involved, as they frequently are, they are relaxed and incontinent. Pain is nearly always present, is of a stabbing character, and is referred to a root area where, as a rule, it is ultimately replaced by anesthesia. It must be remembered, however, that there is sometimes a stage in which pain and anesthesia are co-existent in the same area. Modifications in the sensibility of the affected parts follow the rules indicated in lesions of the peripheral nervous system, and do not conform to those of the spinal cord as laid down by Head and Thompson. Additional assistance in locating the lesion in the cauda equina may sometimes be derived from physical signs pointing to disease of the second lumbar or lower vertebræ.

## II. THE DIFFERENTIATION BETWEEN THE INTRA- AND EXTRA-MEDULLARY ORIGIN OF FOCAL DISEASE.

It not infrequently occurs that, the level of a particular spinal lesion having been diagnosed, it is necessary to come to a conclusion as to whether the lesion is produced by a morbid process originating within or without the cord. The decision of this question may, for instance, have an important bearing upon treatment and may indicate or contra-indicate the performance of a laminectomy.

The early incidence of *pain*, and particularly root pain, to the characters of which reference has been made, is strong evidence of the extramedullary origin of the disease. This kind of pain may be increased by movement, and then suggests some involvement of the vertebral column, or it may be associated with local *tenderness* on pressure and even with *deformity*, which point in the same direction. *Hyperesthesia* in the cutaneous root area, followed by *anesthesia* of a dissociative type, is also found in extramedullary lesions in their early stages as the result of injury to a posterior spinal root.

On the other hand, an *atrophic paralysis* of the muscles innervated from one or two segments preceding spastic paralysis at lower levels, raises suspicions of a primary intramedullary disease, especially if pain has been insignificant or absent during its development. In addition to these differences in local effects, an intramedullary may be distinguished from an extramedullary lesion in some instances by its remote effects. When symptoms below the level of the lesion point for a long time to the *unilaterality* of the latter, an intramedullary cause must be regarded as the more likely. Pressure from without may and does cause unilateral

symptoms, but usually for a short time only, as the effect of compression can, naturally, not long be limited to one side of the spinal cord. Early *sensory* loss, especially of a dissociative type, in regions innervated from below the level of the lesion, favors the intrinsic origin of the disease, and syringomyelia affords the most striking example of this condition.

With regard to the action of the *sphincters* no definite rules can be laid down. Intramedullary disease, even of the cervical region, may sometimes produce bladder troubles before any disturbance of motion or sensation, but this is not a common phenomenon. With extramedullary disease the vesical and rectal symptoms are usually first noticed when the paralysis of one side is being followed by that of the other, or, in cases where the lesion is bilateral from the beginning, as soon as the legs show signs of weakness. In difficult cases skiagraphy may throw some light on the question at issue.

In conclusion, it must be admitted that none of these rules are without exceptions, and that in some instances there is insufficient evidence to give a definite opinion without the aid of laminectomy.

### COMPRESSION OF THE SPINAL CORD.

The possible causes of compression paraplegia are numerous. Fractures, dislocations and other injuries of the vertebral column belong to surgery rather than to medicine, and will not be dealt with here. Inasmuch, however, as the lesion produced by such injuries is of the focal transverse type, its symptomatology at any level can be readily deduced.

The following are the chief causes of compression paraplegia: (1) Fracture dislocation and injuries of the vertebral column. (2) Spinal caries. (3) Newgrowths of (a) the vertebral column and surrounding tissues; (b) of the meninges; and (c) of the cord or its roots. (4) Aneurism of the aorta. (5) Parasites, particularly hydatids in the extrathecal space. (6) Arthritis deformans of the spinal column.

**Spinal Caries or Pott's Disease.—Etiology.**—The tubercle bacillus is necessarily the most important factor, and spinal caries is often associated with tuberculosis of other organs, of which pulmonary tuberculosis is one of the most common. Tuberculosis of the vertebræ occurs most frequently in children and young adults. It may exist at birth, and has been seen in the fetus *in utero*. Injury is often said to precede the onset, and it is possible that a traumatic lesion may determine the site of infection or excite a latent focus.

**Pathology.**—The favorite site of origin for tuberculous disease of the vertebræ is in the spongy substance of the bodies, especially in their anterior halves. Occasionally the first focus is in the cancellous tissue of the laminae or, still more rarely, in the smaller articulations of the spinal column. From its origin in the body of the vertebra there is a tendency to invade the neighboring intervertebral cartilages and vertebræ and to extend posteriorly through the bone and periosteum, so as to invade the extradural space. Having thus reached a region of very limited resistance and considerable vascularity, fungoid granulations spread upward and downward as well as laterally, and finally may attack,

and even, in rare cases, penetrate, the dura mater. If an abscess has formed within the bone, it tends to force its way backward or forward. In the latter case the pus will find a path along the anterior surface of the column beneath the anterior common ligament, and may give rise eventually to a psoas, a retropharyngeal, or other form of "gravity" abscess. In the former case it encounters the posterior common ligament, on one or both sides of which it projects into the lumen of the neural canal. In other instances the pus may find its way into the perispinal tissues, and present itself near the surface in the form of a lumbar abscess.

It is the rule to find some degree of bony deformity; the collapse of one or more diseased bodies produces generally a kyphosis, a kyphoscoliosis, or, occasionally, only a lateral curve. It is obvious that a narrowing of the lumen of the canal may be occasioned (1) by the ingrowth of fungoid granulation tissue, (2) by the intrusion of an abscess, or (3) by displacement of bone. As a matter of fact, more than one of these causes may be at work at the same time. In those rare cases in which an acute compression of the cord is induced, either spontaneously or as the result of some slight injury, it is usually dependent upon a sudden bony displacement still further narrowing a canal which has already been largely encroached upon by granulation tissue.

The injury to the cord is not, however, always a result of compression or of compression alone. According to Schmaus, the most important factor bringing about a loss of conductivity is an œdema of the medullary tissue. This is a consequence of the strangulation of bloodvessels, especially veins, in the surrounding granulation tissue. In the first place the chief effect is upon the myelin sheaths, but later, if the condition persists, the axis cylinders and nerve cells degenerate, and ultimately become destroyed and replaced by proliferated neuroglia. This is the process which earned at one time the name of compression myelitis, but which is more appropriately referred to passive congestion and œdema. Less commonly interference with the arterial supply to a segment or segments of the cord invokes a true evascularization and ischemia, and still more rarely the tuberculous process penetrates the dura and sets up a tuberculous meningomyelitis. The latter may remain localized, but occasionally a general tuberculous meningitis follows. Very uncommonly a tuberculous intramedullary neoplasm may be discovered post-mortem in the neighborhood of the caries.

Dry caries, or *caries sicca*, is the name used to denote that form of the disease in which a rarefying osteitis, rather than abscess or granulations, is the most conspicuous feature. The condition usually proves to be an infective myelitis or meningomyelitis rather than true compression.

The macroscopic changes in spinal caries are very variable, and the cord, in particular, may be found in widely differing states. In one case it will be severely compressed, reduced to a thin ribbon consisting mainly of thickened membranes and sclerosed tissue. In another the caliber may be less diminished, but the consistence, that of cream, indicates the necrotic results of circulatory interference. In another there may be no evidence of compression, but an area, embracing a segment or two, in which swelling and œdema are the most conspicuous

features. Lastly, there are the instances of acute compression, displaying the bruising, softening, and hemorrhage usually associated with fracture dislocations of the vertebral column. Before alluding to the secondary changes it may be mentioned that central cavities of the cord, extending some distance beyond the site of the focal lesion, have been recorded.

The spinal roots may be, and very frequently are, directly involved in the granulation tissue. On the other hand, the anterior roots may be atrophied as a result of the changes taking place in the central gray matter. Ascending and descending tracts of degeneration can be traced above and below the focus of disease in the cord, if it is of sufficiently long standing, and resemble those found in other transverse lesions of the same organ. Not infrequently a diffuse spinal caries may lead to compression at more than one level, in which case the resulting degenerations are all the more complicated.

**Symptoms.**—Caries of the spine is most common in the dorsal, less common in the lumbar, and comparatively rare in the cervical vertebrae and sacrum. On the other hand, it is probable that paralytic cord symptoms are more often associated with cervical than with lumbar caries. In the majority of cases the first symptoms are those which are referable to the disease in the bones, although the results of pressure upon a spinal root, or upon the cord itself, do occasionally manifest themselves before the process in the vertebral column has been suspected. *Localized pain* referred to one part of the spine is usually the first symptom, and has certain characteristics. It is of a dull, boring kind, and is accentuated by movement, especially by pressure applied deeply and directly to spinous processes at its seat. It may also be increased by lateral movements exerted on the part involved, or by jarring the column as a whole. *Localized rigidity* of the spine next deserves attention. In early cases this is due to inhibition of painful movements, and is induced by a subconscious muscular fixation of the part. In later cases mechanical conditions due to displacement of bone or involvement of ligaments may take a share in the production of immobility. The rigidity may be apparent in every movement of the patient, especially when the cervical spine is the seat of disease, or only when he attempts to stoop. In other cases it is only observed in the course of a careful examination.

Next in importance is the presence of a *deformity*. An acute angular curvature denotes a less extensive involvement of vertebrae than a more rounded kyphosis. The most prominent spinous process belongs usually to the vertebra immediately below that which has collapsed. The deformity varies from a slight prominence of one spinous process to a malformation in which the greater part of the vertebral column is directly or indirectly involved. A certain amount of scoliosis is a common accompaniment of, or sequel to, the kyphotic curve, and is often only of secondary origin. On the other hand, scoliosis may be the only form of curvature, in which case it will be distinguished from those of muscular or rachitic origin by the presence of some rigidity and by the failure to reduce it on stooping. Congenital malformations are only to be excluded by the use of skiagraphy. Deformities may be completely lacking, even in those cases of spinal caries which are associated with marked

symptoms of paraplegia, but severe root pains should suggest the possibility of vertebral disease in spite of the absence of all curvatures.

When a spinal root is involved certain symptoms are wont to appear. Of these, the most important are *root pains*, which must be carefully distinguished from the localized pains of bony origin. A root pain is generally sharp or stabbing in character, or may only amount to a constant sense of pressure or tightness. In the case of the limbs the pain is often described as shooting into the periphery; in the case of the trunk it may give rise to one form of girdle sensation. If any *anesthesia* is present as the result of destruction of a single posterior root, it is of narrow limits, and affects the painful and thermal sensibility rather than the tactile. It is very unusual for this to be detected in the trunk areas. On the other hand, an eruption of herpes zoster may suggest that a spinal root ganglion has not escaped injury. When the anterior root-fibres are involved, some *atrophic paresis* of certain muscles may be noted, but this again is slight when only one root is affected, and may be overlooked altogether on the trunk.

In the usual order of clinical sequence, the root symptoms are followed by those which are referable to the spinal compression or to those lesions of the spinal substance which are at any rate induced by the surrounding disease. These are identical with those of any focal lesion of the cord, and are fully described elsewhere, for instance, in the discussion of transverse myelitis. The signs vary with the level of disease, and are referable in the first place to the destruction of certain spinal centres at the site of the lesion, and in the second place to the interference with the conductivity of efferent and afferent tracts. It is impossible to distinguish the signs of destruction of spinal centres from those of the destruction of corresponding roots, and these have already been described. In the case of cervical and upper or middorsal caries a varying degree of spastic paralysis is found below the level of the spinal compression, with a certain amount of sensory loss and of sphincter trouble.

Since the compression is usually of gradual onset, it is well to point out that the earliest evidence is usually to be detected in the examination of the lower extremities. The patient may complain of being easily tired in walking or of a tendency to drag one or both feet, and the observer will find slight weakness of ankle dorsiflexion, brisk knee- and ankle-jerks, and extensor plantar responses. These early signs may precede any anesthesia, and may or may not be associated with precipitate or hesitating micturition. When compression destroys all conductivity in the cord, either after a long time, or as the result of a sudden collapse in rare cases, a flaccid paraplegia will result, and then the deep and superficial reflexes also disappear. Toneless relaxation of the sphincters will also eventuate, as well as complete loss of all sensibility as high as the segmental area corresponding to the spinal lesion.

When the compression involves the lumbosacral region of the cord, a mixture of atrophic palsy of certain muscles, with spastic paresis of others, forms the clinical picture on the motor side. The sensory disturbance follows the distribution of the lumbosacral roots, and the vesical and rectal difficulties are those which are associated with paralysis of

their respective sphincters. The preservation of the patellar and Achilles tendon jerks depends on the integrity of the third and fourth lumbar segments on the one hand and of the fifth lumbar and first sacral segments on the other. The plantar reflex is only obtainable if the lesion extends no lower than the first sacral segment, but is of the extensor type.

Trophic or vasomotor disturbances are common when the spinal lesion is severe, at whatever level it is situated. Sexual impotence results from all but the slightest degrees of compression, and priapism is occasionally observed in the dorsal and cervical cases, with more or less complete paraplegia. Compression of, or above, the fourth cervical segment interferes not only with the intercostal, but also with the diaphragmatic respiration, and lesions of the upper dorsal cord are frequently responsible for distension of the hollow abdominal viscera as the result of splanchnic palsy.

Some reference should be made to the mode of onset and the course of the spinal symptoms. Except in rare cases in which a sudden luxation causes severe bruising or destruction of the cord, the onset of paralysis is slow, taking many days, more often weeks or months, to reach its height. The advent of paralysis may, on the one hand, rapidly succeed the symptoms of vertebral disease, or, on the other, may take place after the latter have been in existence for years. It may be said that no case of spinal caries, of whatever duration, is free from the risk of compression paraplegia.

*Pyrexia* of slight degree, with daily variations, is often present, but the disease sometimes runs an afebrile course, or rapidly becomes afebrile under rest in bed and other appropriate treatment.

The *course* must necessarily vary with the treatment, the nature of the morbid process underlying the spinal lesion, and the severity of the damage, but the condition must always be regarded as chronic, one which is slow to develop and slow to disappear, even when the issue is favorable. Remissions and exacerbations are well known to occur, and may have no obvious relation to the progress of the bone disease. A trauma may induce a rapid accentuation of paralysis, or the spontaneous evacuation of an abscess may be associated with marked improvement. When a favorable turn is taken, the sensory disturbances usually disappear before the motor. In the unfavorable cases death may result from decubitus, cystitis, pyonephritis, or from tuberculosis of other organs. An intermediate course generally means a chronic spastic paraplegia, the length of life depending largely upon the circumstances and surroundings of the patient. The occurrence of gravity abscesses is of surgical rather than medical interest, but it is well to recollect that a psoas or lumbar abscess may be the first indication of vertebral disease, and may indicate the origin of an otherwise unexplained paraplegia.

**Diagnosis.**—From the point of view of diagnosis cases of spinal caries may be divided into two classes: (1) Those in which spinal deformity is present, and (2) those in which the spinal column displays no obvious signs of disease.

1. When the curvature is extensive and prominent the diagnosis is usually easy and straightforward, but slight kyphotic curves are not

uncommon in vertebral carcinoma and aortic aneurism. Prominence of a spinous process is, moreover, occasionally met with in intravertebral tumor originating in the meninges or extrathecal space.

Vertebral *carcinoma* is suspected when malignant disease is, or has been, present in other organs, especially in the breast. In such cases pain, although severe, is less often limited to one particular spot, and examination of the spinal column will probably reveal a loss of the normal curves and a general flatness, which may be combined with some diminution in the patient's height. The age must be taken into account when either carcinoma or aneurism is being considered, and in the latter disease assistance may be forthcoming in the shape of the signs of intrathoracic or abdominal disease. A skiagram may be of the greatest value.

The presence of a scoliosis without kyphosis does not exclude caries, but other causes must be eliminated. The reduction of the curve on extending the spine in the act of stooping, and the absence of rigidity in carrying this out, point to the common forms of scoliosis seen in children and young adults. In such cases there is no paralysis, but a well-marked scoliosis may be seen in syringomyelia, and be associated with spastic paraplegia. The characteristic sensory and trophic disturbances of syringomyelia will serve, as a rule, to differentiate it. Congenital abnormalities of the vertebral column may simulate spinal caries, but a skiagram indicates the true condition.

2. When no spinal deformity is present the inquiry is much more difficult. In the first place, it must be definitely ascertained that the lesion is a focal one and that there are no signs of any disease above a particular level. Having decided that the entire motor and sensory disturbance can be referred to a single focal lesion, and having thus excluded all the diffuse or disseminate scleroses of the cord, the diagnosis will rest between spinal caries, the various forms of intra- and extramedullary tumors, and other, more rare, causes of spinal compression. The differentiation between these conditions must depend, not upon the character and extent of the paralytic phenomena, which may be seen in any one of them, but upon considerations of a wider and accessory character. Tenderness and rigidity of the spinal column at a level corresponding to the signs of compression will, in young children, forcibly suggest vertebral caries. In other persons the same symptoms may be found in connection with vertebral or intravertebral tumors as well as with caries, whether it be of the moist or dry variety. A gumma or local syphilitic meningomyelitis must also be excluded. Here, again, the use of skiagraphy may clinch the diagnosis. It is usually stated that the root pains are more severe and precede by a longer period signs of compression in cases of tumor than in cases of spinal caries, but this is not the universal rule. On the other hand, paralysis more often commences unilaterally when a tumor is the cause of compression than when spinal caries is at work. The detection of tuberculosis in other organs must favor the presumption of spinal caries, but in a considerable number of cases the certain diagnosis must be arrived at by an *exploratory laminectomy*. This should not be delayed too long in doubtful cases, and it is still more important to remember that the diagnosis of a chronic myelitis

to explain slow progressive signs of paraplegia is never admissible except in certain rare syphilitic cases, which can generally be excluded by the results of appropriate treatment or by examining the cerebrospinal fluid.

The pain complained of by neurasthenic patients is often referred to the spine, which may be exquisitely tender. It is rare, however, for the pain and tenderness to be constantly localized at a particular level, and examination generally shows that the tenderness is more superficial than deep. Local rigidity is not observed in these cases, although the patient may hold the spinal column stiffly as a whole.

The differentiation between a true compression paraplegia and a *functional paraplegia* may sometimes require attention. In the functional cases the legs are usually stiff and extended, without tendency to flexor spasms. The weakness is general throughout all the groups of muscles instead of being more marked in the dorsiflexors of the ankles and flexors of the knees, as it is in all cases of spastic paraplegia of organic origin. The characteristic interference with the sphincters is absent in the functional cases, and, although the tendon jerks are generally exaggerated and a form of clonus of irregular rhythm may be elicited, the plantar responses are either absent or flexor in type. That these points are of importance is evident when the possibility of a functional paraplegia supervening in a spinal caries is remembered.

**Prognosis.**—There are few general rules which can guide one in giving a prognosis in any particular case of spinal caries which is complicated by paraplegia. Youth favors the outlook and age adds to the gravity. Oppenheim gives statistics from Billroth's clinic which show that out of 97 patients suffering from caries, 48 died, 22 were cured, and 11 were dismissed as incurable. Speaking generally, the more complete the paralysis the more serious is the case as regards life, on account of the complications which arise in connection with decubitus, cystitis, etc. It may be admitted that the cervical cases are more dangerous, on account of the interference with the respiratory musculature, than many of those in which the mid- or lower dorsal region is involved, but the writer has been favorably impressed by the power of recovery exhibited by some cervical cases, especially in young subjects. Tuberculosis in other parts naturally adds to the danger, but treatment may have as good results upon one focus as upon another, and the presence of pulmonary tuberculosis, for instance, is not an insuperable obstacle to recovery. Gowers truly says that there is no disease of the cord in which symptoms of equal gravity so often pass away.

**Treatment.**—This is a question which may provoke considerable diversity of opinion, especially in regard to the advantages and disadvantages of operative interference.

*Preparalytic Prophylaxis.*—The earlier the disease of the vertebrae is recognized and treated, the less likely are cord symptoms to supervene. This is an axiom of importance, but it is not equivalent to a statement that paraplegia will not develop in cases which have had the advantage of early and good treatment. As soon as caries of the spine has been diagnosed the patient should be placed on his back, kept at complete



rest for many months, and the general treatment for tuberculosis should be carried out.

*Rest and Extension.*—In certain cases of spinal caries in which paraplegia has supervened, especially in young children, the adoption of simple rest will suffice to restore power to the paralyzed limbs in a comparatively short length of time. In others recovery will ensue after many months, even when no other remedial measures are superadded. The application of extension to the vertebral column may, and often does, make all the difference in those cases in which rest alone has proved of no avail. Extension should certainly be tried in all cases where displacement of bone is obvious and recent, but it is often useless and adds to the discomfort of the patient when the bony deformity is of long standing. It is probably more serviceable in cervical than in dorsal caries, and is of little value when the lumbar spine is the seat of disease. In all cases in which rest alone or rest and extension are being used every precaution must be taken against bedsores.

The employment of plaster, poroplastic, and leathern jackets cannot satisfactorily replace recumbency in bed, but is necessary as a supplementary measure in order to maintain immobility for a time after recovery has taken place. In very young children the use of bandages and splints may be necessary to insure the requisite rest. Massage, passive movements, and perhaps electricity may be requisitioned while the patient is in bed in order to improve and maintain the nutrition of the paralyzed limbs, so long as no undue movement is imparted to the trunk.

*Operative Treatment.*—This should be regarded as a supplementary measure, one not to be undertaken lightly and not to be regarded as replacing rest and open-air treatment. It may be resorted to before rest has been tried, when there are indications that no relief of pressure can be procured by the latter, or it may be employed when a course of rest has failed to produce any amelioration in the paraplegia. The evacuation of any superficial abscess must be carried out at once, and a deep abscess, if within reach, should be subjected to the same treatment, particularly when it is suspected of causing compression symptoms. On the other hand, a deep-seated abscess may sometimes dry up without surgical interference under favorable conditions. Laminectomy should not be postponed until secondary changes in the spinal cord render the relief of pressure ineffectual in restoring power to the paralyzed parts, and, speaking generally, it should be undertaken earlier in adults than in children.

It is probable that surgical interference would enjoy a greater popularity if the cases were chosen with greater care and submitted to the surgeons at an earlier period. The indications and contra-indications for laminectomy as usually cited are not to be relied upon. For instance, it is often stated that the involvement of several vertebrae excludes the use of the knife. The writer has had an experience which directly contravenes such a view. A child, aged three years, with cervical caries, after five months of rest, during which period symptoms of paraplegia continued to progress, was operated on by D. Armour. A large abscess was evacuated, and what appeared to be the necrosed remains of several vertebral

bodies were extracted. The abscess cavity was washed out and the wound closed. Within four months the child, with a shortened neck, was running about without a sign of paralysis and without any need of artificial support for her head.

In spite of the success which attends operation in some suitable cases, it must be remembered that spinal caries is not a condition with regard to which it may be said that an operation can do no harm. There are instances in which this course has appeared to excite the disease into renewed activity, and others in which a fatal issue from tuberculous meningitis has rapidly followed. It may justifiably be hoped that the use of skiagraphy will encourage and promote that careful selection of cases which is necessary to bring this treatment into the position it deserves. Equally important is it that emphasis should be laid on the after treatment, so that each may enjoy for a prolonged period the rest, fresh air, and good nourishment which are essential for satisfactory results. Finally, no hesitation should be felt in advising laminectomy when the patient's life is seriously threatened by increasing respiratory embarrassment, as is not infrequently the case in caries of the higher spinal regions.

### TUMORS OF THE SPINAL CORD.

For the sake of convenience, although at the expense of terminological accuracy, the title "Tumors of the Spinal Cord" is here, as often elsewhere, intended to include all tumors which habitually modify the functions of the spinal cord by pressure upon or invasion of that organ, as well as those which originate in its substance. The classification of spinal tumors followed by Bruns may be adopted here, but it is necessary to explain that the word tumor embraces, in this connection, granulomata and parasitic cysts as well as neoplasms proper. On the other hand, spinal gliomatosis is reserved for the article on syringomyelia.

I. Tumors which, originating in its envelopes, secondarily affect the spinal cord.

(a) Vertebral tumors arising from the spinal column or the soft tissues immediately surrounding it.

(b) Intravertebral tumors, which may be divided into two classes in accordance with their relation to the dura mater.

1. Extradural tumors originating in the periosteum of the vertebræ, the outer layers of the dura mater, or the fatty areolar tissue of the epidural space.

2. Intradural tumors originating from the inner layers of the dura mater, the arachnoid, the ligamentum denticulatum, the spinal roots, or the pia mater.

II. Intramedullary tumors of intrinsic spinal origin.

**Vertebral Tumors.**—The bones of the spinal column, especially their bodies and less commonly their arches and processes, are frequently the site of malignant growths.

*Carcinoma* in this region is always secondary and generally metastatic, although occasionally the vertebræ are directly involved by extension of

disease from other organs. The primary focus may be in the uterus, stomach, intestine, prostate, thyroid, lung, kidney, or gall-bladder, but most commonly, by far, is in the mammary gland. This special connection with mammary carcinoma and perhaps uterine cancer is sufficient to account for the greater incidence of the disease in females, although males are not exempt. Probably 70 or 80 per cent. of cases occur in women. The age which predisposes to vertebral carcinoma is that of carcinoma of other organs. Considering the rarity of bronchial carcinoma, secondary vertebral disease from this source is relatively frequent.

The lumbar and dorsal parts of the column are more often involved than the cervical, and the disease may be limited to one or more foci or spread diffusely over many vertebræ. It is not uncommon to find the spine much more extensively affected after death than was suspected during life. The neoplasm is found to attack first the spongy portions of the bone, to destroy the medulla, and to fill the medullary spaces. The more compact bony cortex may survive as a thin shell for a time, but is ultimately also replaced by the expanding growth. Along with osseous resorption, a certain amount of calcification in portions of the neoplasm is not infrequently observed. When this osteoplastic process is excessive a compact and firm ankylosis of a large part of the vertebral column may ensue, and there may be a complete absence of any osseous deformity. On the other hand, it is more common for the softening produced by the infiltration of a spongy growth to allow the body weight to effect a general shortening of the column, a condition which is sometimes described as "entassement." When the softening is local it is often described as carcinomatous caries, and, like the tuberculous variety, may lead to collapse and the formation of a curve like that of Pott's disease. In this manner the spinal cord may be compressed, but is only affected in comparatively few cases, the spinal roots being much more prone to suffer close to or within the intervertebral foramina. Nodules of carcinoma projecting from the inner surface of the vertebral bodies or arches may exert pressure upon the cord, and very occasionally the tissue of the epidural space becomes invaded by growth, with the result that the cord is surrounded by a kind of neoplastic cylinder. Very rarely does the disease penetrate the theca.

*Sarcoma* of the vertebral column or of the soft tissues immediately surrounding it may be primary or secondary, may form a single well-defined tumor, or be diffused as multiple growths. The lumbar and sacral regions appear to be the favorite seat of the disease. The results of the infiltration of the vertebral column resemble those of the carcinomata, and may comprise collapse and luxation, the formation of curvatures, and the compression of nerve roots or of the spinal cord. The spinal cord compression is usually brought about by the presence of growths in the epidural space, but occasionally by the penetration of the dura along the course of a spinal root, and the formation of masses upon the surface of the pia. The *secondary* sarcomata of the spinal column either extend directly from the neighboring tissues or are metastatic deposits from more distant parts. The writer has seen a renal sarcoma invade the vertebral column along many of the spinal nerves,

and evoke symptoms both of root involvement and of compression paraplegia. Metastatic sarcoma is a rare growth in the spinal bones; according to Schlesinger secondary invasion of the vertebral column is most common when the primary sarcoma is of osseous origin.

*Myeloma* occupies an intermediate position between the malignant and benign tumors of the vertebrae. Multiple myelomatosis is usually characterized by a wide infiltration of the bony tissue of many vertebrae, rendering large extents of the column soft and pliable, with the result that deformities are easily produced by the influence of weight or position. Less often one or more masses of growth are found in the form of definite tumors. Other bones, especially the ribs, sternum, clavicle, etc., are frequently involved at the same time. The spinal cord may suffer from compression, in consequence of deformities leading to narrowing of the neural canal or from a nodule of growth projecting from the internal surface of a vertebra. Myelomata are not invariably soft throughout; occasionally they indulge in ossification and productions of ivory-like character. The disease is confined to the osseous system, and affects males more often than females.

The *benign tumors* of the vertebral column are very rare, and comprise osteomata and exostoses, chondromata and osteochondromata, myxomata, the so-called luxuriant callus formations, and the bony excrescences associated with arthritis deformans. The bony tumors only affect the nervous system when they grow in such a way as to encroach upon the neural canal or the intervertebral foramina. The cartilaginous growths may penetrate the intervertebral foramina and produce root palsies and sensory symptoms. The occasional erosion of the spine by aneurism of the aorta or by hydatid cysts may be mentioned, but it is still an open question whether the latter ever originate primarily from the bony envelopes of the spinal cord.

**Intravertebral Tumors.**—1. **Extradural.**—Tumors of the epidural space are more often secondary than primary. Sarcomata and lipomata share the reputation of being the most common varieties, but fibromata, myxomata, chondromata, are also very occasionally seen in this situation. It is only rarely that the malignant growths penetrate the dura mater. Hydatid cysts are found most commonly in this region when they give rise to spinal symptoms.

2. **Intradural.**—These originate from the inner layers of the dura mater, the arachnoid, the ligamentum denticulatum, the spinal roots, or the pia mater. Sarcomata may be diffuse or localized. The former variety, which may be termed pial sarcomatosis, is usually of the round-celled type, and characterized by its tendency to envelop more or less extensive portions of the spinal cord in a kind of cylindrical covering. It is a growth of the leptomeninges, and is usually less luxuriant on the ventral than on the dorsal and lateral aspects of the cord. Not infrequently it may cover the whole length of the cord and cauda equina and penetrate even into the cranial cavity. In some instances it never actually invades the substance of the cord; in others it infiltrates that organ as well as the spinal roots. In spite of the absence of direct invasion by the neoplasm the cord is often softened owing to the interference with its vascular

supply and perhaps to some degree of pressure. More rarely secondary growths from sarcomata of the vertebrae, the pleura, or peritoneum gain access to the subdural space and extend in a longitudinal direction upon the surface of the cord. Localized circumscribed sarcomata are also found in this situation and tend to exert mechanical pressure upon the spinal marrow as well as upon spinal roots. Endotheliomata arising from the theca, cylindromata (Billroth), psammomata, fibromata, lipomata (Gowers), lymphangiomata (Schlesinger) are less common. On the other hand, fibromyxomata or fibrosarcomata are frequently found in connection with the nerve roots. These are often single, but neurofibromata are generally multiple, and may be associated with more or less neurofibromatosis of the peripheral nervous system.

Such vascular growths as angiosarcoma and true angioma are observed now and then within the dura, and cysticerci as well as echinococci are also recorded as present in the same cavity. Intradural tumors are usually situated laterally or posterolaterally, rarely on the anterior surface of the cord. The latter is often displaced to one side and compressed or indented. In the region of the cauda equina there is room for greater development in lateral dimensions than in the higher regions, and neoplasms may here reach a considerable size with relatively few symptoms.

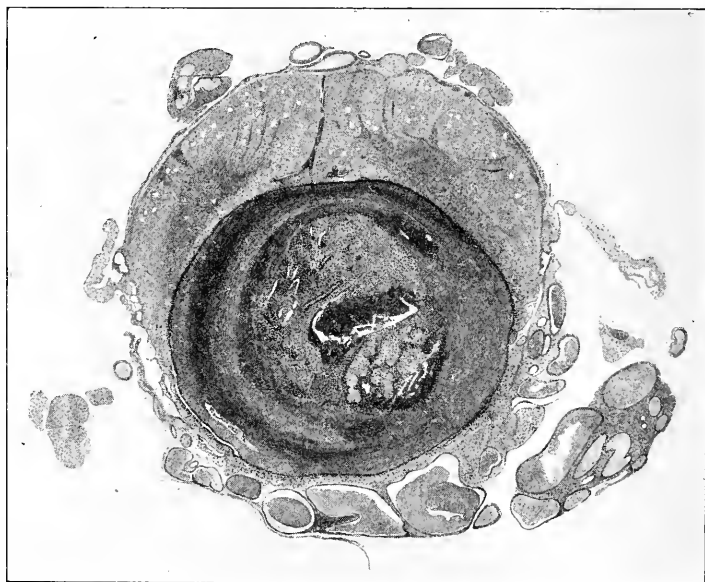
**3. Intramedullary Tumors.**—These comprise gliomata, sarcomata, angiosarcomata, as well as tubercles and gummata. Cysticerci are rarely found in the substance of the cord. Gliomata are usually diffuse and conform to the type which is discussed under syringomyelia, but they are occasionally circumscribed, producing the ordinary changes and symptoms of an intramedullary growth. The sarcomata, solitary tubercles, and gummata arise in the first instance from the pial septa, and are more often single than multiple. Cases in which gummata have been found in the brain, in the cord, and on several spinal roots, are described.

**Pathological Processes in the Nervous Tissues Resulting from the Pressure of Spinal Tumors.**—Meningeal tumors rarely occasion more than slight erosion of the bony parts, but their effects upon the nervous structures within the vertebral column are important. The spinal roots, although more resistant than the spinal cord, are usually the first to suffer. They may be directly involved in the growth, in which it is sometimes difficult to trace their course, or they may be compressed as they pass through the dura or even the intervertebral foramina. Degenerative changes follow in either case, and may be traced in the efferent fibres in their peripheral, and in the afferent fibres in their intramedullary course.

The spinal cord may suffer in different ways. In most cases a certain amount of displacement is the first result, and this is followed by compression. The early effects of the latter may be only mechanical, but sooner or later they lead to interference with the circulation and the production of œdema and ischemia. Venous stasis has usually been regarded as an important element in producing the œdema, and the arterial and capillary blood supply is modified in consequence of the latter. Superficial arteries may also be compressed, thus contributing to the local anæmia. The more specialized nervous structures, particularly the ganglion cells, are the

first to show signs of nutritional change, and the myelin sheaths of the conducting tracts readily undergo degeneration. More pronounced degrees of ischemia lead to areas of softening, which may be associated with sclerotic changes in the neuroglia if the pressure is slowly exerted. The presence of secondary degeneration above and below the site of compression denotes definite changes in the axis cylinders at that level. Similarly degeneration of the anterior root fibres follows the atrophy of the cells of the ventral gray matter. The cord may be softened without direct compression, owing to the interference with the arteries, veins, and lymphatics on the surface. Direct infiltration of the cord by extramedullary tumors is less frequent than might be supposed, the pia-arach-

FIG. 15



Gumma of third lumbar segment.

noid affording a certain amount of protection. Intramedullary growths may be sharply defined or may present little in the way of differentiation from the nervous matter. Those which are circumscribed are often surrounded by an area of softening, and occasionally by evidences of inflammation or of toxic oedema.

**Incidence.**—Schlesinger has shown that tumors affecting the cord are by no means common, and says that in 35,000 necropsies only 147 instances were recorded.

**Age.**—Starr collected 100 cases of intravertebral tumor, and found that 70 occurred above and 30 under the age of fifteen years. The following table represents the age incidence in 82 cases collected by Armour from the records of the National Hospital (London).

Year.	Vertebral carcinoma.	Vertebral sarcoma.	Spinal tumors.	Medul- lary.	Intra- dural.	Extra- dural.	Both.
Under 10 . . .	0	0	1	1	0	0	0
11 to 20 . . .	0	4	10	2	3	4	1
21 to 30 . . .	0	1	11	3	5	3	0
31 to 40 . . .	1	3	12	4	5	3	0
41 to 50 . . .	6	1	17	2	10	4	1
51 to 60 . . .	2	2	5	1	4	0	0
61 to 70 . . .	3	1	1	0	1	0	0
71 to 80 . . .	1	0	0	0	0	0	0
Total . . .	13	12	57	13	28	14	2

**Nature and Site.**—Of 400 cases collected by Schlesinger, 126 were intramedullary and 239 extramedullary. The extramedullary included 151 intradural and 88 extradural growths.

	Medullary.	Intradural.	Extradural.	Both.
Cervical . . . . .	5	5	0	1
Cervicodorsal . . . . .	0	1	1	0
Dorsal . . . . .	4	13	10	1
Dorsolumbar . . . . .	0	1	2	0
Lumbar . . . . .	3	2	0	0
Cauda equina . . . . .	0	6	1	0
Sacrum . . . . .	0	0	0	0
Diffuse . . . . .	1	0	0	0
Total . . . . .	13	28	14	2

**Symptoms.**—These can be conveniently divided into three groups. The first comprises those associated with disease of the bone, the second those which indicate involvement of the spinal roots, and the third those referable to derangement of the functions of the spinal cord.

**Vertebral Symptoms.**—The earliest symptom is *pain* referred to some part of the back, dull, boring, or burning in character, and liable to exacerbations, either spontaneous or the result of pressure, movement, or concussion. Conscious of the increased suffering, which is excited by movements of the trunk or head, and particularly by those which involve rotation of the vertebral axis, the patient holds himself stiffly and avoids exposing himself, as far as he is able, to the possibility of a sudden jar. On examination the observer will detect limitation of the normal mobility of one or more parts of the spine, and will easily elicit evidences of tenderness in the same regions. Inspection and palpation of the back and even of the anterior surface of the sacrum by rectum may fail to reveal any mass, especially in carcinoma. On the other hand, obliteration of the normal spinal curves, shortening of the vertebral column, or difficulty in distinguishing the spinous processes by palpation, may afford indications. The most striking examples of loss of height are seen in vertebral carcinoma or myeloma, and may be inferred from the statement of the patient or from the obvious collapse and condensation of certain parts of the column. A narrowing of the normal space intervening between the lower ribs and the iliac crests, and the resulting prominence given to the abdomen, may afford some indication of this “*entassement*.” While obliteration of natural curves is marked in some cases, the formation of abnormal kyphoses is equally characteristic of

others. A sharp-angled curvature is generally acquired, more or less rapidly, when the body of a vertebra becomes so softened as no longer to be able to bear the weight of those above. More rounded kyphosis is the result of a general and diffuse infiltration such as is seen in cases of multiple myelomata.

The degree of prominence to which bone symptoms attain in any particular case varies considerably. On the one hand, pain and tenderness of the back, deformities, and curvatures may monopolize attention throughout the course. On the other hand, pain and paresthesia referred to the extremities and disturbance of general health may fail to attract particular notice to the spinal column, and very extensive disease of that part of the skeleton runs the risk of escaping detection.

A word is necessary upon the occurrence of vertebral symptoms in instances of *intravertebral* tumor in which the bones escaped involvement. It is not rare to observe limitation of movement, some local tenderness, and even the prominence of a spinous process beyond its neighbors; the occurrence of these physical signs must not be allowed to weigh too heavily against the diagnosis of a meningeal growth.

**Spinal Root Symptoms.**—It is desirable to utter a word of warning against placing the symptoms and signs of the spinal tumors derived from this source in too important a position. From a positive point of view their occurrence is valuable, but it is unwise to hold that the absence of any phenomena associated with disease of the spinal roots in any way negatives the existence of a spinal tumor. *Pain* is by far the most common of root symptoms, and yet a painless or almost painless development of paraplegia is met with sufficiently often in intravertebral neoplasm to prove that this symptom has sometimes been held in too high esteem.

The pain associated with disease of the posterior roots, whether in their intrathecal or intravertebral course, is usually severe, neuralgic, stabbing, and referred to the peripheral distribution of the afferent fibres. It may be unilateral or bilateral, and in the latter case may give rise to a painful form of girdle sensation. Exacerbations may be caused by movement, especially when the root is involved by growth in the intervertebral foramen, which is most frequent in vertebral carcinoma or sarcoma. The skin of the areas in which pain is felt is often hyperesthetic, and later may become anesthetic and analgesic, although pain persists (*anesthesia dolorosa*). Appreciable anesthesia and analgesia are rarely evident unless two or more consecutive roots are interfered with, although some loss of thermal and painful sensibility may be detected in the case of single root lesions in connection with the upper or lower limbs. When the spinal root ganglion is involved an eruption of herpes zoster may appear in the corresponding root area.

The destruction of the anterior root fibres gives rise to physical signs which are difficult to separate from those of compression of the cord itself, and which often make their appearance at a time when the latter organ has begun to suffer. Thus, abolition of function in a single anterior root does not, as a rule, give rise to complete palsy or complete atrophy of any muscle, but a few muscles, partially innervated by the root involved, present some paresis, some wasting, partial reaction of degen-



eration, and possibly fibrillary twitchings such as are seen in cases of progressive muscular atrophy. These signs are much more readily detected in the limbs than in the trunk, where the overlapping of muscular root innervation is much more pronounced. When two, three, or more adjacent anterior roots are affected, the paralysis and wasting is more or less complete in certain muscles and partial in others, the contractures usually associated with lower motor neuron palsy following in the ordinary course of events. It is hardly necessary to add that the reflexes subserved by the affected spinal roots are abolished. Cases of tumor of the cauda equina afford the most striking examples of multiple root lesions uncomplicated by symptoms of spinal cord origin.

**Spinal Cord Symptoms.**—Sooner or later the neoplasm gives rise to the symptoms of a focal lesion of the spinal cord. In vertebral carcinoma death may occur before any compression has taken place, but in the majority of instances of intravertebral growth the spinal symptoms engross most attention before the disease has been very long in progress. It is characteristic of spinal tumors that their effects upon the cord are progressive, more characteristic, indeed, than the occurrence of pain. The symptoms attendant upon transverse lesions and the diagnosis of their level have already been fully discussed, but a few points, bearing upon the onset and course of spinal tumor, must be referred to here.

With vertebral tumors signs of compression are mostly bilateral from the beginning, with intravertebral and particularly intradural growths, although they are frequently situated to one side of the cord, the symptoms of paralysis are rarely unilateral for long. On the other hand, inequality in degree of disablement of the two sides is common enough, and it is not always the side first affected which is later the more profoundly paralyzed. True unilateral lesions of considerable duration are more commonly met with in cases of intramedullary tumors, and their symptomatology is that of Brown-Séquard's paralysis.

In *extramedullary* growths motion is usually affected before sensation, and the earliest symptoms of compression paraplegia in cases of tumor above the lumbosacral enlargement are easily induced fatigue in walking and a tendency to drag the toes. Examination at this time will probably reveal some paresis in the dorsiflexors of the ankle and flexors of the knees, as well as increased tendon jerks and the extensor type of plantar reflex. These signs may, however, only be found on one side at first. So long as the symptoms are unilateral there is rarely any interference with the sphincters, but the extension of the effects of compression to the other side is attended by precipitancy or hesitancy in micturition. Increasing spasticity and paresis in the muscles innervated from the segments below the site of disease mark the advance of pressure until a time comes when the lesion amounts to a complete physiological section; a flaccid paralysis then replaces the spasticity, tendon and superficial reflexes disappear, and the sphincters become quite incontinent.

Similarly, on the *sensory* side paresthesia in the periphery of the lower extremities—a vague numbness or coldness of the feet—precedes the development of definite diminution of sensibility. When the latter is established it involves all forms of sensibility, perhaps in unequal degree,

and spreads rapidly upward toward the level of the disease. For some time the upper limit of diminished sensibility may correspond to a segmental area some distance below that of the site of compression, the decussation of tactile, painful, and thermal impulses being gradual, and their supradecussational paths lying more exposed to the effects of pressure than the more central areas in which the crossing takes place. It is common, however, to find that, while diminution in cutaneous sensibility only reaches a certain level, between that level and the area innervated from the segment which is being compressed, stimuli, tactile or painful, may be felt by the patient as altered in character, although they are perfectly appreciated. The more nearly the lesion approaches completeness, the more closely does the loss of sensibility approximate upward to the area subserved by the segment which is on the verge of physiological disintegration. Emphasis is laid upon this fact, because it is often of supreme importance to estimate accurately the upper limit of sensory change in order to localize the site of compression. In other cases, in which the afferent root is compressed, or the posterior gray matter suffers early at the level of the tumor, the local anesthesia in the corresponding segmental area is sufficient to guide the observer to a correct estimation of the height of the lesion.

Sensory loss in cases of *intramedullary* growth tends to exhibit special features. Its early appearance, its frequent unilateral distribution, and dissociative characters are among the most important of these. The sensory phenomena associated with Brown-Séquard's paralysis have been described, but a duplication of these may sometimes be caused by a central tumor, with the result that the local and remote areas of dissociative anesthesia are met with on each side of the body at the same time. Although this is characteristic of syringomyelia, it is not limited to that disease, and may occasionally be found with medullary tumors other than gliomata.

**Upper Cervical Region.**—Tumors in this region tend to produce a picture of spinal hemiplegia or double hemiplegia which resembles in many respects that of pontine or cerebral origin. The absence of symptoms pointing to involvement of the cranial nerves and the local signs in the region of the neck usually separate the two conditions. In these cases pain is referred to the neck and to the region of the occiput, extending as high as the vertex, and is followed by the development of hemiplegia involving the arm, leg, and trunk on one side, less commonly by bilateral motor paralysis. In a case seen by the writer, pressure, exerted by a tumor at the level of the foramen magnum, caused hemiplegia on one side, shortly followed by a similar condition on the other, the side last affected being the more disabled at the time the patient came under observation. Although usually regarded as a dangerous site, in relation to life, it is worthy of note that the patient in this instance had suffered for over three years from symptoms referable to the presence of the growth. More profound paralysis of the respiratory musculature would have brought about an early fatal termination.

**Cervical Enlargement.**—Owing to our fairly precise knowledge of the spinal motor and sensory innervation of the upper extremities, tumors

of the cervical enlargement can generally be readily recognized. An atrophic paralysis of the arm, with spastic paralysis of the parts below, associated with sensory disturbances and sphincter troubles, all steadily progressive, are characteristic. Oculopupillary symptoms and grave respiratory paresis may often be noted in the same connection.

**The Dorsal Region.**—The chief difference between tumors of the dorsal cord and those of other levels lies in the absence of well-marked atrophic palsies as local symptoms at the level of the lesion. This is partly due to the overlapping of the muscular spinal innervation, but still more to the difficulty in distinguishing clinically how far separate sections of a large muscle supplied by many spinal segments are in a state of functional activity, or how large a share of a movement, such as inspiration, is carried out by each separate muscle, such as an intercostal, concerned in it. These difficulties are not so great if a careful examination is made, and there are certain points which are helpful. Paralysis of the lower half of the rectus abdominis is readily demonstrated by the upward movement of the umbilicus in the attempt to rise from a supine to a sitting posture. Palsy of the lower intercostals may be recognized if the hands of the observer are placed over the lower ribs during respiration, especially if the patient is not too well covered with fat. Perhaps the accurate testing of the superficial trunk reflexes from segment to segment affords more dependable evidence of the level of the lesion than any of the motor phenomena, but the accomplishment of this requires considerable experience and is often difficult in obese subjects. If, for instance, the skin reflexes are obtained on either side above the umbilicus and are absent below that level, the indication is in favor of a lesion reaching as high as the tenth dorsal segment, and this may be confirmed by the movement of the umbilicus already referred to. On the whole, the disturbances of sensibility offer, in most cases of tumors of the dorsal cord, the most reliable indication of the height to which the disease has attained. Apart from the questions involved in their segmental localization, tumors of the dorsal cord present no special clinical features if exception is made for the splanchnic palsy, occasionally associated with pressure on the upper half of this region. The distension of the abdomen in these cases is often only temporary, but, while it lasts, may be productive of great discomfort and respiratory embarrassment, as well as of troublesome constipation. The sphincter troubles and the tendency to decubitus depend directly on the severity of the lesion and the amount of interference with the sensory tracts.

**The Lumbosacral Enlargement.**—The short length of this part of the spinal cord and the fact that it can be easily exposed by the removal of two or three laminae minimizes the necessity for the accurate segmental localization which is so necessary in cases of tumor of other regions. On the other hand, it is most important to distinguish between lesions of this enlargement and those of the cauda equina.

Pains and atrophic muscular paralysis of the regions innervated by the lumbar plexus, sensory disturbances over the whole of the lower extremities, absence of patellar jerks and presence of ankle clonus and extensor responses, together with retention of urine and feces, form a symptom

complex characteristic of compression of the lumbar segments and roots. When the lesion also involves the sacral segments, the atrophic paralysis and anesthesia are more extensive, and absence of all reflexes in the legs is combined with incontinence of the sphincters. Under these circumstances the ultimate picture may closely resemble that of a tumor of the cauda equina, and the diagnosis may depend upon a consideration of the development of the symptoms. A Brown-Séquard paralysis is occasionally met with in cases of lumbar tumor, more often with intra- than extramedullary growths.

**The Cauda Equina.**—It must be admitted that tumors, apart from injuries, form the large majority of morbid conditions of the cauda equina. The first symptoms of some neoplasm attacking the lumbosacral roots is nearly always pain in one lower limb. The supervention of anesthesia and of atrophic muscular palsy and the gradual spread of these three signs in that order, from one part to another, together with the early involvement of the opposite side in a more or less symmetrical manner, point with certainty to a gross organic basis for the disease. The loss of sphincter control when the third and fourth sacral roots begin to suffer, and the abolition of the various reflexes, with the spread of the morbid process to the radicular paths, upon the integrity of which they depend, follow in the natural course of events. The question as to whether the growth is of vertebral or intravertebral origin is sometimes difficult to answer, but the solution may possibly be arrived at by a careful examination of the lower lumbar vertebræ and sacrum, supplemented by investigation of the pelvis.

Contra-indications to the diagnosis of tumor of the cauda equina are afforded by the presence of a Brown-Séquard's paralysis, of any spasticity, exaggeration of tendon reflexes or extensor type of plantar reflex. It may be mentioned here that neither the presence nor absence of fibrillary tremors, nor the character of the electrical reactions of the muscles, can be relied upon for the purpose of distinguishing between an affection of the anterior gray matter and of the anterior roots.

**Course and Result.**—With few exceptions the course of spinal tumors is gradually progressive and their result fatal. The rapidity with which symptoms reach their greatest intensity differs considerably with the nature and seat of the growth, and the relative duration of the pre-paralytic and paralytic stages may present wide variations. Rare instances of apoplectiform paraplegia are afforded by the collapse of diseased portions of the spinal column and by the development of acute "myelitic" changes in the cord at the seat of slight pressure, but in the first cases the morbid condition of the bones has usually made itself obvious by other symptoms, and in the others premonitory signs of compression have afforded some clue to the presence of a tumor, even if local or root pains have been absent.

With regard to duration a sharp distinction may be drawn between cases of vertebral and intravertebral growths. In the former, whether of carcinomatous or sarcomatous origin, the inevitable fatal issue is generally reached within nine months; in the latter, life may be prolonged for two or two and one-half years, and in some instances for much longer

periods. Isolated nodules of sarcoma, fibroma, or psammoma have been known to produce symptoms over eight, fourteen, and even twenty-five years. Remissions and exacerbations cannot be said to be marked or even usual, but, they are not by any means of great rarity.

The primary intravertebral tumors are hardly ever responsible for metastases in other parts; it is less uncommon for intracranial growths to be associated with secondary deposits in the spinal cord or on the spinal roots.

The mortality depends on the almost inevitable supervention of decubitus, cystitis, and renal disease, except in those cases in which respiratory paralysis leads to a rapid death.

**Diagnosis.**—Three important questions have to be answered in this connection. In the *first* place, the differentiation between a spinal tumor and some other spinal or nervous diseases has to be established. In the *second* place, the precise situation of the neoplasm must be ascertained. *Thirdly*, the nature of the growth is a matter of some moment when the line of treatment has to be decided upon.

1. **The Diagnosis of Spinal Tumor.**—This is not so difficult a task in the case of *vertebral* tumors, because, on the one hand, these are frequently secondary, and the presence or history of another growth may indicate the nature of the disease, and, on the other, spinal caries is almost the only morbid process with which they may be confounded, if an exception is made for vertebral aneurism and hydatids. No absolute rules can be laid down, but in a general way caries of the spine is associated with more marked curvatures and gibbosities, with less prolonged, less severe, and perhaps more symmetrical root pains, and often with definite skiagraphic appearances. Vertebral tumors are characterized by less sharp curvatures and more condensation of the spine, by prolonged and severe pains, often asymmetrical, and by a more rapidly progressive course and cachexia.

Tuberculosis in other organs must always be looked for. In their early stages vertebral tumors are often overlooked because pain may be the only symptom and physical examination may yield no further help. To neurasthenia or "rheumatism" must not be too hastily assigned persistent pains in patients who are obviously suffering, but it is not often that a definite decision can be arrived at until some more objective evidence is available.

With *intravertebral* growths there are certain diagnostic points of primary importance. The fact that all symptoms can be ascribed to a single lesion at a particular level in the cerebrospinal system serves to exclude such diffuse diseases as disseminate sclerosis, in which paraplegia, sensory and sphincter disturbances may have a more or less gradual and painless onset. When everything points to a single localized lesion, the fact that the onset is slow and the course of symptoms progressive practically excludes all forms of spinal disease except a tumor, a local meningeal cyst, and some rare forms of spinal syphilis. With the exception of the latter there is no form of chronic myelitis which can simulate the insidious onset or the progressive course of a compression paraplegia. The rare cases of vertebral caries without deformities or evidence of

osseous disease must be remembered, although it is often impossible to diagnose these without exploration. Mention has just been made of local meningeal cysts with the object of emphasizing the difficulty of their differentiation from spinal tumors, this being the greater on account of our ignorance of their pathogenesis. That they are a real and not infrequent source of error is proved, although the mistake is of academical rather than practical or therapeutic importance.

*Hypertrophic cervical pachymeningitis* may suggest the early stages of an intravertebral neoplasm, but the limitation of the symptoms to those of root or meningeal origin points in the direction of the true diagnosis, which may often be confirmed by the favorable course.

As a matter of fact, there are few conditions with which a spinal tumor can be confounded if we exclude caries and meningeal cysts, and if we remember that a tumor produces a local lesion and that the onset of symptoms is slow and their course steadily progressive.

When multiple tumors of the meninges or roots exist, there is usually much difficulty in forming a true estimate of the amount and situation of the spinal damage. Multiple neurofibromata and multiple sarcomata, although located on the roots, often evoke symptoms of spinal cord compression before giving rise to anything very definite in the way of radicular palsies or disturbances of sensation. The symptom complex may be so mixed in degree and height that a true conception of the anatomical condition is impossible.

2. The exact situation of the tumor must be gauged from the data which have been given, but attention may be directed to certain general principles. In estimating the highest level of damaged spinal marrow the opinion must be arrived at by a careful comparison of the results of examination on the motor, sensory, and reflex sides, helped by any information which can be gained from investigating the spine of the patient manually, visually, or by the x-rays. The most minute changes in sensibility should be noted, especially in case of tumor of the dorsal region, and when a segmental area shows well-marked anesthesia it should be concluded that at least one spinal segment above that by which the area is innervated is, in all probability, severely damaged. Thus, if definite anesthesia can be traced as high as the seventh dorsal segmental area, the sixth dorsal segment is compressed, and the removal of the fourth and fifth dorsal laminae will expose the seat of disease. The highest indication of any disturbance of function is nearly always the safest guide to follow. The downward extent of the lesion is generally impossible to estimate.

3. It is hardly ever possible even to hazard a guess as to the histological characters of a primary intravertebral tumor until it has been exposed. It is therefore unwise to allow the question of its nature to influence in any way the consideration of treatment. On the other hand, a vertebral tumor, primary or secondary, or a secondary intravertebral growth, usually presents evidences or associations pointing to its nature. In the great majority of cases the knowledge thus acquired contra-indicates the hope of a radical cure.

The diagnosis between extra- and intramedullary lesions of the cord

has been discussed, and the principles there laid down can be applied to cases of spinal tumor. It is necessary to repeat the warning that there are not infrequent instances in which the question can only be positively answered by an appeal to exploration, and that the gravity of the outlook justifies that measure in any doubtful case.

**Prognosis.**—In the large majority of cases of vertebral tumors their malignant character renders the prognosis hopeless and the fatal termination a matter of months. With intravertebral growths the outlook depends entirely on the possibility of successful removal, the liability to recurrence and the condition of the spinal cord at the time the attempt to remove them is made. The prognosis must always be grave until these questions can be answered. The most favorable cases are those of localized benign or comparatively benign tumors in the extradural or intradural spaces, and the least favorable those of diffuse or multiple extramedullary neoplasms and all those of intramedullary origin. It is fortunate that compression of the cord may be prolonged and severe without destroying all hope that its functional activity can be restored if the source of pressure is removed.

**Treatment.**—From the therapeutic point of view cases of spinal tumor may be divided into operable and inoperable classes. With those which are inoperable remedial measures must be directed toward the alleviation of pain and the prevention of complications, such as sepsis, bronchitis, pneumonia, cystitis, and pyonephritis. The alleviation of pain may be difficult, but less compunction need be felt about the administration of narcotics than in diseases with a more favorable outlook. Unfortunately the pain produced by pressure upon the afferent roots and the great discomfort evoked by flexor spasms of the lower limbs in the stage of advanced spastic paraplegia do not always yield even to opium. In this contingency the advisability of dividing the posterior roots for the relief of the more urgent sufferings may be well considered.

The possibility of a syphilitic basis for the symptoms of spinal tumor, in the shape of an intramedullary gumma or a local gummatous meningitis, must not be forgotten, and antisyphilitic remedies should always be given a trial in doubtful cases. The examination of the spinal fluid should be resorted to for the purpose of setting these doubts at rest.

A large number of cases of intravertebral tumor are suitable for surgical exploration, even if only a small proportion attain a really successful result. With our present knowledge it is usually possible to give an accurate diagnosis of the site of the lesion, but rarely possible to state the nature, size, origin or possibility of removal of the growth. Nothing short of exposure of the tumor can clear up these points, and there is less harm done by operating on cases of what prove to be ineradicable spinal neoplasms than by choosing unsuitable cases of intracranial growth for surgical intervention. An opportunity is afforded for relieving pain even when the disease cannot be arrested, and the operation in skilled hands is attended by no undue risk considering the gravity of the case. The surgeon who can stay his hand when a laminectomy exposes a hopeless condition will do no harm even if he effects no good.

This is not the place to discuss surgical details, but one or two points

may be noted. It sometimes happens that on the removal of two or three laminae no tumor is found in the expected situation. The absence of pulsation in the theca at this level should encourage the surgeon to go higher, and he will probably find the source of pressure by removing another arch or two. It is less common for the laminectomy to be performed at too high a level, but in such a case the healthy pulsation of the theca may suggest the advisability of extending the wound downward. The removal of several arches is not attended by the disastrous results in the way of lack of support which might be expected.

The after treatment of successful operations is that of convalescing myelitis. The restoration of function in paralyzed limbs may be hastened by the employment of massage and electricity, but attention to the general nutrition and health of the patient will do the rest.

### CAVITIES AND FISSURES OF THE SPINAL CORD.

Under this title are included a number of conditions of little practical importance with the exception of syringomyelia. For some reason we are accustomed to term "cavities" in the spinal cord what would, perhaps more properly, be termed "cysts" in other organs. At any rate we mean by a cavity a space, with or without an organized wall, containing fluid. Cavities, continuing the abuse of this term, are the not infrequent terminal results of an acute inflammation, in the course of which portions of the spinal tissue have become necrosed and ultimately absorbed. An example may be found in the cord of a long-standing case of poliomyelitis, just as a cyst or porencephaly may represent an old encephalitis. Of very similar origin are the cysts or cavities resulting from an old focal hemorrhage or thrombosis. These cavities have no definite lining membranes, their walls consisting of neuroglia, which may or may not be compressed, so as to give it a condensed laminated appearance.

The term "fissure" is usually applied to a cavity which is narrow, and which may appear narrower after it has lost its fluid contents than it did before it was opened. Care must be taken not to regard as fissures those spaces which are seen in microscopic specimens as the result of hardening processes, and which are particularly liable to be artificially formed along the tracts of bloodvessels when the walls have been thickened by disease or when the embedding has not been skillfully carried out.

**Hydromyelia.**—This name has been variously used, and it would cause less confusion if it were confined to cases of simple dilatation of the central canal without co-existing alterations in surrounding structures, apart from the results of simple stretching, and without clinical symptoms. The condition is most frequently found in infants with congenital hydrocephalus, spina bifida, or other developmental abnormalities, but may also be present unsuspected in older persons in minor degrees. A satisfactory explanation of its production is difficult, but increase of fluid pressure due to mechanical causes, or physiological increase of fluid output by an overactive ependyma, may be of sufficient account in certain cases. The hydromyelic cavity is lined by ependymal cells, and its shape is usually round, but sometimes T-shaped, owing to a



diverticulum in the direction of the posterior median septum. The addition of some peri-ependymal gliosis or of ependymal proliferation with the formation of accessory canals is the first step toward the production of what is termed "syringomyelia," and is evidence of the very imperfect line of demarcation separating the two conditions.

**Syringomyelia.**—**Definition.**—A chronic disease of the spinal cord characterized anatomically by the existence of one or more pathological cavities, and clinically, in its typical form, by the presence of dissociative anesthesia and trophic changes in muscles, skin, and bone, together with numerous other, but less constant, physical signs. It should be stated that Morvan's disease, first described in 1883, and for a time regarded as a separate disease, is included in this article only as a variety of syringomyelia. This is justified by the prevailing opinion that there are no grounds for putting it in a distinct category.

**Etiology.**—**Distribution.**—Syringomyelia can nowhere be regarded as a common disease, although it is now well recognized in all civilized nations. There is no reason to suppose that it is not universal in its distribution, and yet its literature appears to indicate that it is of more frequent occurrence in France and Germany than in America or the British Isles. Statistics are wanting, and it may be that the more careful investigation of the infirmity class of patients in the first-named countries accounts for the apparent difference in numbers.

**Sex.**—It is usually stated that men are more frequently affected, and this is borne out by an analysis of 40 cases which have been in the National Hospital (London). Of these, 25 were men and 15 women.

**Age.**—The second and third decades represent the most frequent time of onset. In the 40 cases just mentioned the first indication of any abnormality was observed on an average, at the age of twenty-four and one-half years. Among the males the average age was twenty-eight and among the females nineteen, a difference which would probably be less marked in a larger series. The latest date of onset (fifty-six) was in a man, and the earliest in a girl of seven.

**Heredity.**—There is no evidence that heredity plays any important part, and, although its occurrence in more than one member of a family has been recorded, it cannot be termed a familial disease.

**Congenital Anomalies.**—The not uncommon presence of congenital anomalies in the victims of syringomyelia is important. A careful study will often reveal some skeletal peculiarities. The patient may be unduly small, much smaller than his brothers and sisters; he may be infantile in proportion, presenting a large head with small trunk and limbs. Another patient may have unusual deformities of the skull; for instance, bosses in the occipital or temporal regions; he may have, without other signs of acromegaly, very large hands and feet. Pronounced degrees of genu valgum or of pes cavus have been present from early infancy in cases of the disease, and examination of the spinal column may elicit signs of a spina bifida occulta.

**Trauma.**—A history of preceding injury must be regarded with judicious suspicion in all diseases, and syringomyelia is no exception. It is safe only to say that more or less severe trauma to the head and spine

is by no means an infrequent antecedent, and that considerable importance has been attached to this fact. The relationship of spinal hemorrhage to syringomyelia is intimately connected with this question.

*Other Diseases.*—Syphilis may be excluded as a direct cause, but its influence in producing meningitis and vascular disease must be taken into account. The common infective diseases do not appear to have any causal relationship, although the occurrence of one or other of them may coincide with the development of, or may intensify, its early symptoms. The co-existence of conditions such as tabes, spinal caries, etc., is rare and probably unimportant; on the other hand, cases of combined acromegaly and syringomyelia have been recorded sufficiently often to make the association interesting and worthy of further investigation.

*Pathology.*—The exposure of the spinal cord is generally sufficient to reveal characteristic changes. The organ is altered, often very irregularly, in shape; the cervical region, as a rule, is enlarged and flattened, the lateral dimension being proportionately more increased than the antero-posterior. The dorsal segments share in the change, or may be narrow with irregular thickenings. Usually the lumbosacral enlargement has a more normal appearance. The extension upward of the cervical deformity may cease high up in the cord, may occasionally involve the medulla, or very rarely the pons. Folds and furrows are not infrequently observed on the surface of the cord, especially if the fluid contents have drained away. The meninges, in the majority of cases, present no abnormality. Exceptions are found in those cases of spinal cavitation definitely associated with pachy- or leptomeningitis (often syphilitic in origin), a considerable number of which have been recorded. The anterior spinal roots, particularly those of the cervical enlargement, may be atrophied and of a gray translucent color. On palpation the swollen parts are soft and usually fluctuate; the narrower regions may be normal in consistence or suggest the presence of a hard core lying within an outer ring of the medullary substance. A series of transverse slices will demonstrate striking changes in the interior of the cord, probably varying considerably at different levels. In the region of the greatest swelling, usually the cervicodorsal region, the section may at first sight appear to have traversed an area of necrotic softening, or even an abscess, but closer scrutiny will show that near the centre of the cord is a cavity, from which clear fluid may be oozing, and that this cavity is surrounded by an enveloping mass of gelatinous material either pale and translucent or yellowish brown, according to the amount of altered blood pigment it contains. The immediate wall of the cavity sometimes stands out as a more opaque pale yellow membrane which has been thrown into folds by the escape of the contents and collapse of surrounding parts. Further sections at different levels will discover the length of the cavity. In some cases it extends through a few segments only, in others through the whole length of the cord. Rarely the tube is prolonged into the fourth ventricle, although in the unique case described by Spiller it reached the right internal capsule.

Frequently the cavity disappears at one level, the surrounding opaque tissue continuing for another segment or two, sometimes to develop

another cavity beyond. Two cavities may be present side by side, but if these are traced farther it is usual to find that one is an offshoot of the other. The lumen varies both in size and shape at different levels.

*Position of Cavity.*—To describe this accurately it is necessary first of all to know its shape and size at any particular level, but speaking generally, these tubes lie behind the anterior commissure, their centre of origin, if one may use the expression, being situated in the gray matter behind the central canal, either on the median line or laterally in the base of one or other posterior cornu. From this centre diverticula may extend in any direction, with resulting varieties in the shape of the cavity in transverse section, but more often than not some part of the posterior white columns is invaded at one level or another. Without doubt the posterior horns and posterior columns are more often involved than the anterior horns, while the anterolateral white matter is rarely encroached upon directly by the cavity, although it may suffer indirectly from the effects of pressure. The shape of the cavity may be round, oval, crescentic, stellate, or irregular, and it is certainly unusual to find any great degree of symmetry in its relation to the two halves of the cord.

In the bulb, as in the cord, the morbid process has a favorite site. Occasionally the spinal cavity may reach the fourth ventricle, but more often it ends below, the only evidence of the disease in the bulb being a fissure or fissures originating in the floor of the ventricle, a little to one side of the midline, and extending forward and outward in such a way as to cut off the restiform body from the central parts of the medulla. Such a fissure may destroy the descending root of the fifth nerve, the solitary fasciculus and some of the nuclei belonging to the vagoglossopharyngeal nerves, but it rarely extends as high as the facial nucleus. It is sometimes unilateral and sometimes bilateral.

What has been said with regard to the position of the spinal cavity applies equally to the disposition of its surrounding tissue, which roughly follows the various diverticula when it does not form prolongations of its own, and which, it must be remembered, extends in most cases to levels beyond the ends of the tube.

Deformities of the surface of the cord and of the outline of the gray and white matter are dependent upon, and present as many varieties as, the distortions of the cavity and its walls.

*Gliomatosis.*—The glial tissue surrounding the cavity varies enormously in thickness not only in different cases, but at different levels in the same case. It may only present the characters of a thin lining membrane to a large cavity, or it may in other instances be so voluminous as to overshadow completely the narrow slit-like cavity it contains. In fact, it may exist in the form of a hard core without cavitation through considerable lengths of the cord. Whatever may be the prevailing theory as to its mode of production, there can be no doubt that it is essentially of neuroglial origin, the term gliomatosis being useful in that it suggests an overgrowth of that tissue and yet separates it from the true glioma which belongs to the group of malignant neoplasms. This gelatinous-looking material is mainly composed of fibres and cells whose relative proportion varies, but it may be stated roughly that there are more cells than in

normal neuroglia, and less cells than in a true glioma. At levels where no true cavity exists, foci of softening, to which the name granular disintegration has been applied, are occasionally present in the centre of the gliomatous formation.

*Central Canal.*—The fact that the cavity may be lined or partly lined by ependymal cells has been noted, and this condition usually obtains when the cavity represents an enlarged central canal or when a cavity formed in the peri-ependymal tissue has fused with the central canal. Rarely, a cavity situated at some little distance from an intact central canal may be lined with ependymal cells, in which case the explanation is offered that the cavity has originated in a nest of embryonal tissue persisting from the time of the closing up of the posterior columns. In most cases the cavity is either obviously a dilatation of the central canal or appears to be quite independent of the latter. According to Schlesinger the independence is only apparent. At any rate it is quite a common thing to see the central canal, either open or obliterated, lying quite separate from the pathological cavity in a long series of sections. In other instances the cavity is fused with the central canal over several segments and separated from it in regions beyond.

*Vascular Lesions.*—In a certain number of cases the vessels of the cord and its membranes are perfectly healthy, but in others the vessels have undergone definite alterations. For this reason French observers in particular have attached considerable importance to “les espaces vasculo-conjunctifs.” Vessels radiating from the surface of the cord into the central glioma present the following changes: The vessel walls and the accompanying glial tissue are swollen, hyaline, and often thrown into folds. As a rule, the lumen is diminished or obliterated, especially when it reaches the glial mass, and there is no evidence of proliferation of the nuclear elements of the walls or sheath. In the neighborhood of the cavity the only remnant of a vessel may be an undulating ribbon of hyaline connective tissue which has been unfortunately called a papillary membrane, and this, owing to degeneration of neighboring tissues, may at times line the cavity or a portion of its wall. Schlesinger has observed these vascular changes in a case of syringomyelia without gliomatosis.

In sections the central parts of the glial mass are usually poor in vessels, but the more peripheral zones contain a number, mostly running parallel with the long axis of the cord. It is stated that newly formed vessels are found in these parts, but the evidence of this does not appear to be very clear. Hemorrhages old and recent are by no means infrequent.

*Lesions of the Nervous Elements.*—The gliomatosis and cavitation, after attaining a certain bulk, produce changes in the surrounding tissues partly by compression and partly by direct invasion. The latter method is the one by which the posterior columns are principally affected, the newly formed neuroglia eating its way, as it were, between the bundles and producing slow destruction. Closely associated with compression is the production of an œdema of the tissues surrounding the glial mass which is responsible for the rarefaction of the gray matter often seen in the anterior horns. The nerve cells become isolated and their processes disappear, with the result that a species of cavity may be formed to

which the name perigliomatous, as opposed to endogliomatous, has been given. A still further loss of glial tissue may eventually lead to fusion of the cavities originating in these two different ways, and thus to the appearance of a large central canal surrounded only by a thin layer of white matter.

The secondary degenerations which result from the lesions of the white matter and from the destruction of ganglion cells in the gray matter conform to the general laws governing secondary degeneration in the cord and peripheral nerves. Sclerosis of the pyramidal tracts, of the posterior columns, and of the ascending anterolateral tracts is of common occurrence, as well as atrophy of the anterior roots.

Mention must be made of certain neuromata which have been described in the gliomatous tissue, white matter, intramedullary roots, and subglial tissue. These small tumors are composed of numerous fine medullated fibres running in various directions and accompanied only in some instances by bloodvessels. Their origin is uncertain, Raymond suggesting that they afford evidence of attempted regeneration, and Schlesinger that they are due to local irritation. Patoir and Raveirt have found neurofibromata in the spinal roots in a case of syringomyelia, and have regarded them as of similar origin to the neuromata of the cord and perhaps allied in their etiology to the interstitial changes occurring in the peripheral nerves. The pathological changes which occur in other tissues, in the nerves, muscles, skin, joints, and bones, must be all considered as secondary to the lesions of the central nervous system. They do not present features peculiar to this disease.

The occurrence of syringomyelic cavities in association with marked pachymeningitis or leptomenigitis or with vertebral caries is not extremely rare, although the exact relationship to the spinal condition has not been fully established. Still more common is the concurrence of some true neoplasm with the syringomyelia. At any level of the cord, commonly in the cervical region, may be found a true tumor, involving more or less the entire transverse area, which has evidently originated from a part of the gliomatous mass. Such are commonly very cellular gliomata or very vascular angiogliomata, but may sometimes be of a more sarcomatous nature. The writer has seen neoplasms of this kind in the medulla, the cervical and sacral regions in different cases, all associated with syringomyelia.

**Pathogenesis.**—Numerous theories have been put forward to explain the origin and progression of the morbid process. Their number suggests that many cases of spinal cavitation and gliomatosis, although resembling one another roughly in their clinical and anatomical characters, may yet have different modes of origin, and this view is finding more and more favor with those whose knowledge of the disease is most profound. While it is impossible to elaborate dogmatically a genetic theory applicable to every case, a consideration of certain developmental, anatomical, and pathological facts cannot fail to throw some light on particular examples, and may serve as a basis for more scientific grouping.

**Developmental.**—The medullary canal of early fetal life is only represented clearly in the child by the central canal of the cord, a cylindrical

tube running throughout the length of that organ lined by ependymal cells of epiblastic origin. The central canal, however, represents only the anterior limb of the medullary canal, the posterior and lateral limbs disappearing in the coalescence of the posterior columns of either side along the posterior median fissures and in the process of forming the gray commissure and the bases of the posterior horns. Normally no cells resembling the ependymal cells of the central canal can be seen in the gray commissure, in the bases of the posterior horns, or in the walls of the posterior median fissure. Presumably, however, there are in these three contiguous parts embryonic remnants, which may take on renewed activity and once again produce neuroglia, the most primitive tissue of epiblastic origin in the central nervous system. On such an assumption the occurrence of a neuroglial hyperplasia appears to have a reasonable, if not complete, explanation and gliomatosis practically always originates either in the gray commissure, the bases of the posterior horns, or in the anterior third of the posterior columns. The occasional presence of groups of ependymal cells away from the central canal may be regarded as a developmental accident in otherwise normal cords, but it is possible that such groups may be the starting point of a periependymal hyperplasia, just as the latter certainly originates in some instances from the walls of the central canal itself.

*Anatomical.*—A consideration of the spinal vascular supply will bring out one important point. That part of the spinal cord which includes the gray commissure, the anterior third of the posterior columns, and the bases of the posterior cornua is the centre of the cord from the vascular point of view. It receives blood from various radiating vessels, of which the most important are those entering along the posterior roots and along the posterior median septum, as well as from some of the terminal branches of the anterior spinal artery. It is the central terminus of those various arteries, and contains, therefore, vessels of small caliber, some of which have a transverse and some a longitudinal course.

This is of importance in two ways. In the first place, any general disease of the spinal arteries which reduces the muscularity and elasticity of their walls will produce its greatest effect on the vascularization of this central zone. In the second place, relying as it does very largely on the posterior spinal arterial system for its blood supply, any morbid process leading to pressure upon or strangulation of the pial vessels on the posterior surface of the cord will have a profound effect upon the same area.

The lymphatic supply of this central spinal area must also be considered, the more so because theories suggesting an infective origin for the disease have been propounded. The central canal is possibly an important lymph channel, and the presence in it of an irritant agent capable of inciting ependymal proliferation and peri-ependymal gliosis has been suspected. More important is the fact that lymphatic infection of the cord from other organs generally takes place along the lymph channels of the posterior roots. On facts such as these, and experimental work in connection with lymphatic infections of the spinal cord through the

peripheral nerves, some have attempted to isolate a group of syringomyelias due to an ascending neuritis.

The closure of the central canal by pressure at one level of the cord has been quoted as a sufficient reason to explain dilatation of that canal at other levels. The justice of this is by no means proved, and, in fact, may be questioned when it is remembered that in normal adult cords the canal may be obliterated in some segments and yet patent without dilatation above and below.

There are certain *pathological* facts to be taken into account, of which the most important is the partiality of intramedullary hemorrhages and abscesses for particular paths in their progress through the cord. There is a great tendency for blood and pus to track from segment to segment along the tissues at the bases of the posterior horns or in the central gray matter, so much so that this area has been termed the zone of least resistance. It is evident that gliomatosis spreads along similar lines, and the close relationship existing between the two processes has led to the belief that many cases of syringomyelia have their remote origin in spinal hemorrhage occurring at birth. Cases have been recorded among adults in which the clinical history has suggested a traumatic intramedullary hemorrhage followed, perhaps years later, by the symptoms of a progressive syringomyelic lesion. It is possible, therefore, that an old pathological process may after a lapse of time be the starting point for gliomatosis and cavitation.

There are other forms of syringomyelic cavities apparently originating from a level of the cord, which has been compressed by meningitis, by vertebral caries, or by some other external cause. It is difficult to see how this comes about unless as the result of evascularization of the cord with the production of necrotic cavities in the central zone and the subsequent spread of disintegration along the favorite lines just mentioned. The presence of gliomatous tissue surrounding the cavity so formed is not easy to explain, but the theory has been advanced that the products of disintegration are sufficiently irritating to incite glial proliferation. Such an explanation appears in a few cases more likely than one which assumes the necessary existence of a congenital abnormality as a predisposing factor.

These developmental, anatomical, and pathological data, together with clinical observations, suggest that various influences are at work in the production of spinal cavities. Many examples of syringomyelia fall naturally into groups in which one or other of these influences plays a predominant part, while others are not so easily classified. Thus there are cases in which a congenital defect appears to carry great weight; others in which a morbid condition of the vascular supply seems too important to overlook, and others in which some source for a chronic lymphatic infection or a history of some traumatic hematomyelia require careful consideration. The fact that gliomatosis is always associated in greater or less degree with the formation of these cavities can only be explained on the supposition that the neuroglia of the central zone of the cord has an inherent tendency toward this form of overgrowth, the initiation of which may be due to a variety of agents, physiological,

chemical, mechanical, or pathological. The tendency exhibited by the gliomatous tissues after reaching a certain bulk to undergo central degeneration and then to help in the formation of cavities must be referred either to deficient vascular supply or to some inherent property.

With regard to the ordinary cases of chronic progressive syringomyelia little hesitation can be felt in placing foremost some congenital anomaly of the central embryonal tissue, the latent activity of which is prone to display itself during the early years of adult life in the form of a slow-growing hyperplasia, having some of the characters of a benign neoplasm and strong tendencies toward the formation of cavities. As far as the other morbid influences we have referred to are concerned, they can only be looked upon as secondary exciting agents or, in very occasional instances, as of primary causal importance.

**Symptoms.**—A disease so prolific in physical signs and symptoms as syringomyelia can never be portrayed by a serial enumeration of its manifestations. The detailed consideration of these must be postponed, therefore, until a picture has been drawn of an ordinary typical case which may serve as a basis for further remarks. A man, aged twenty-seven years, seeks advice on account of some paralysis of his hands. He says that the first symptom occurred about four years before, when a sensation of numbness in his left hand was followed before very long by some weakness and awkwardness of the same part. After some months he noticed that the hand was wasting, the tendons and bones standing out more prominently than on the other side. Little notice was taken of this until he became aware that similar alterations were taking place in his right hand and that he was beginning to find himself awkward in writing, buttoning his clothes, and other fine movements. The legs are perhaps more easily tired than formerly, and on one or two occasions the left toes have caught a step in going up stairs. He says that he has had little or no pain, an occasional "burning sensation" in the region of the neck, but nothing to worry about.

The patient appears healthy and strong, and his face is natural, except that the left pupil is smaller and the left palpebral fissure narrower than the right, giving an appearance of slight ptosis on the left side. This, however, disappears on his looking up, both upper eyelids moving equally well. The pupils react to light, but do not dilate very readily to shade, nor do they react when the skin of the neck is pinched. The ocular movements are natural except that lateral deviation is associated with marked nystagmus. The observer is struck by the good muscular development of the shoulder and upper arm compared to the wasted condition of the forearm and hand. This is more marked on the left side, where the contour of the inner border of the forearm is completely altered and flattened. The left hand is noticeable for the "claw" position of its fingers, the flatness of its palm, and for the fact that the palmar surface of the thumb is parallel instead of nearly at right angles to that of the other fingers. There are an unusual number of scars on the hand and forearm, and the patient, on being questioned, admits to many cuts and burns. One long scar was the result of leaning his forearm on an oven door, the only knowledge of its having been burnt being conveyed



to him by the discovery of a blister. The terminal phalanx of the middle finger is missing, and on inquiry it appears that three or four years ago, subsequent to a slight injury, a whitlow formed, which in spite of incision and scraping on two or three occasions ended after many months in the loss of the top of his finger. There was no pain with it. The skin of the hand is rough and hard, and the remaining nails are thick and striated. All his finger movements are feeble, and flexion of the wrist is much less powerful than extension. Except for the triceps, which may be a little impaired, the muscles about the upper arm and shoulder are quite as strong as their appearance would suggest. Turning the man around, the only thing to catch the eye is a slight yet definite lateral curvature in the dorsal part of the vertebral column, of which the patient is quite unaware. Inspection of the legs reveals nothing visibly wrong, although, if each group of muscles is tested against resistance, dorsiflexion of the feet is not so powerfully performed as it should be. A glance at his boots shows that the toes of each are unduly worn down, more noticeably on the left side. Tapping the patellar or Achilles tendon produces brisk jerks, and stimulation of the soles of the feet elicits an extensor response on either side. An attempt to obtain the abdominal reflexes is unsuccessful. The patient's sensibility to tactile stimuli is found to be perfect everywhere, but of painful and thermal stimuli his appreciation is practically absent over a large area, which includes both arms, the neck, and the thorax as far down as the xiphisternum on the left side and the third rib on the right. On the other hand, his sense of passive movement and position and his stereognosis are intact even in the upper limbs.

The subsequent history illustrates the usual course of events. After a temporary improvement in the strength of the muscles of the upper limbs due to local treatment, the inevitable slow progress of the disease displays itself in the fact that several more muscles in each arm are affected and the area of analgesia and thermo-anesthesia extends lower down the trunk and over a great part of the left side of the face. Five years later the patient is found to be unable to work on account of the condition of his hands, and his walking has become seriously interfered with by the increasing spasticity of his lower limbs. The right shoulder has recently become the site of a large fluctuating swelling, a skiagram showing that the joint has become completely disorganized. The left side of the tongue is slightly wasted and the left palatal arch lags behind the right during phonation. Inquiry elicits information that the patient's control over his vesical sphincter is not so good as it was, and hesitating or precipitate micturition is the result. A few more years and the bedridden stage is reached, the length of which will depend very largely on the circumstances surrounding the sufferer and the care with which he is nursed. Some pulmonary complication or septic infection will end an illness which may have lasted anywhere from fifteen to thirty-five or even forty-five years.

This is the usual progress, but there are exceptional cases, in which no increase in symptoms may take place for many years, and some in which the disease appears to become permanently arrested. Slow, insidious

progress is the general rule, although every now and then a case is observed in which a sudden increase in symptoms has been followed by some improvement, leaving the patient, however, worse off than he was before the sudden change. Such an event has generally been attributed to hemorrhage taking place into a syringal cavity.

Such a history is typical of the majority of syringomyelic patients and these may be included in a class to which the name "*amyotrophic*" is applicable on account of the early appearance of progressive muscular atrophy and the prominent part taken by that symptom throughout. The remaining cases may be divided into three other classes, making four altogether, viz.: (1) Amyotrophic, (2) spastic, (3) Morvan's type, and (4) spinal tumor type.

This classification is useful, although quite artificial, and depends upon the predominance of certain clinical features. Thus, the *spastic* case ("forme spasmodique") is characterized by marked rigidity affecting all four limbs and producing attitudes and fixed positions which are worthy of note. The back is kyphotic, the head low between the shoulders and forced forward in a manner suggestive of paralysis agitans. The hands, too, adopt special attitudes, the three inner fingers flexed into the palm, and the index firmly pressed against the thumb. Such a case may and usually does present some muscular atrophy, although the latter is not so striking as in the amyotrophic class, in which spasticity is less marked and usually confined to the trunk and lower extremities.

The *Morvan's type* of case, at one time regarded as a separate disease, is remarkable for trophic changes in the extremities, especially for a slow necrotic dactylitis, in the course of which most of the small bones of the hands or feet may be lost and the parts deformed almost beyond recognition. Amyotrophy and sensory disturbances are associated with the trophic changes in skin and bones.

The fourth class includes a number of cases in which a diagnosis of spinal tumor is more often made than that of syringomyelia until operation or postmortem examination reveals the presence of a cavity associated with a local gliomatous or sarcomatous tumor, to which the symptoms and physical signs have been mainly due. In such cases the absence of the characteristic dissociation of sensation and of trophic disturbances may have rendered an accurate diagnosis difficult or impossible.

*Onset.*—In 17 of 40 cases this was marked by some disturbance of motion or sensation of one of the upper extremities. In at least half of them the patient first noticed some sensation of cold, heat, numbness, tingling, or pain in the hand or forearm, but in all the weakness and subsequent wasting either followed quickly or were the earliest to appear. At the time they came under observation all had some objective loss of sensation in the affected limb, and it may well be that this preceded the other signs, without its existence having come to the patient's notice. Some difficulty in gait was stated by 8 patients to be their first trouble, but it is remarkable how rarely any incoördination in walking is seen later in the disease. Subjective sensations in the lower extremities were noted before any other symptoms in three cases, one of whom

described his right leg as cold and dead, and another his left leg as always hot when the rest of the body and the other leg were cold.

Scoliosis preceded all other signs of disease in four cases, and painless whitlow in a like number. Pain in the occiput and neck (in two instances), difficulty in articulation, and difficulty in micturition were the other initiatory symptoms.

**Disturbances of Motion.**—(a) *Atrophic Paralysis.*—This is more common in the upper extremities than elsewhere, usually beginning in the intrinsic hand muscles on one side and spreading up the arm in accordance with the segmental innervation of the muscles and not following the peripheral nerve distribution. Less often it commences in the muscles of the shoulder girdle and rarely in the muscles of the legs. Awkwardness or feebleness of finger movements usually precedes the appearance of obvious atrophy, but later on the amount of paralysis corresponds roughly to the destruction of muscle fibres. In consequence of the interosseal palsy and the preservation of the long extensors of the fingers the hand may present the “main en griffe” appearance. When vasomotor and trophic changes are associated, the “main succulente” is the result. Any muscle which is undergoing atrophy may display fibrillary tremors, and its response to faradic and galvanic currents is proportionately diminished; in some instances true reaction of degeneration is observed. All atrophic muscles sooner or later lose their tendon reflexes.

(b) *Spastic Paralysis.*—The degree of spastic paralysis depends upon the amount of degeneration in the pyramidal tracts and its extent upon the level at which the pressure upon, or the invasion of, the lateral columns occurs. Mention has already been made of the spastic form in which the affection of the pyramids extends at least as high as the upper cervical segments, with the result that all four limbs as well as the trunk are more or less rigid. In the more common amyotrophic or, speaking anatomically, cervicodorsal form the spasticity usually appears later and affects the trunk and legs only. In addition to their passive and active rigidity, severely spastic parts are subject to flexor spasms, sometimes of a painful character, which may occur spontaneously or as a more or less easily elicited reflex phenomenon. Associated with the spasticity is the form of flexed hand already referred to and a tendency to clubbing of the feet. All spastic muscles have increased tendon reflexes sometimes amounting to clonus. Occasionally a moderate degree of atrophy is associated with spasticity in the same muscle, which may preserve a tendon jerk of diminished excursion and power. More often than not the atrophic paralysis of one hand is accompanied by spastic paresis of the leg on the same side before the opposite limbs have become affected. In all spastic cases it is the rule to find diminution or absence of the abdominal reflexes and an extensor response to plantar stimulation. These constitute the earliest evidences of this form and are present before any definite paresis or rigidity can be detected.

(c) *Incoördination.*—A marked degree is not a common feature, and the majority do not suffer from any great loss of sense of position and active movement. Some interference with this sense may be present in those patients who complain of reeling gait in the early stages. Tabes

and syringomyelia occurring in the same individual is not unknown, but the association is only accidental.

(d) *Spinal Curvature*.—The common dorsal scoliosis must be regarded in most instances as of muscular origin. The asymmetrical character of the paresis affecting the spinal muscles and the legs is quite sufficient to account for its presence in the ordinary way. When the deformity is extreme or when combined with some kyphosis it is possible that osseous or arthritic changes may be additional factors.

(e) *Tremors*.—These are not common features, although fine tremors, resembling those of Graves' disease, and coarse "intention" tremors have been described.

*Bulbar Symptoms*.—When the disease affects the bulb either primarily or as an extension from the cervical region of the spinal cord the clinical picture is often described as "syringobulbia." Atrophy of the tongue, unilateral or bilateral, paralysis of one or both vocal cords or palatal arches are among the more constant symptoms.

Facial paralysis is more rare, and ocular palsies very exceptional. On the other hand, *nystagmus* is a common feature, and this may be lateral, vertical, or rotatory. The latter type was present in 2 out of 40 cases, and the former was of frequent occurrence. Sensory disturbances in the area supplied by the trigeminal nerve are often present, even without other bulbar symptoms. Defects in articulation, deglutition, and phonation are the results of the various palsies mentioned.

The only special senses to be interfered with in any degree of frequency are those of taste and hearing, and even these are usually intact except in severe examples of the bulbar form. Vision remains unaffected almost without exception, but H. S. Hutchinson has once observed degeneration of the optic nerves.

**Disturbances of Sensation.**—(a) *Thermo-anesthesia*.—This is one of the most constant symptoms, and is almost invariably present to some extent even in the earliest stages. The upper extremities or thorax are usually first affected, extension of the area taking place with the progress of the disease. One patch of thermo-anesthesia is sometimes separated for a time from another by a healthy area, coalescence occurring later on. The defect in sensibility exists generally for both cold and heat and for all degrees of temperature, but occasionally cold is recognized and heat is not, or vice versa. It may happen that an area of complete thermo-anesthesia has a border separating it from the normal skin, in which cold only is appreciated. When the defect of sensibility is only partial, it would appear, according to Rosenfeld, that the power of appreciating heat or cold depends on the extent of the area stimulated rather than on the degrees of temperature.

The association of thermo-anesthesia and analgesia together with complete preservation of tactile sensibility in the same cutaneous region is very characteristic, but tactile anesthesia may coincide with the other forms, especially in advanced stages. Although thermal sensibility may be absent from the skin and from the mucous membrane of the mouth and gullet, yet appreciation of the temperature of a liquid may

be derived from the stomach. A syringomyelic patient may burn his mouth and be unaware of the heat of the food until after it is swallowed.

(b) *Analgesia*.—More often than not analgesia is present in thermo-anesthetic areas. Frequently the distribution of the one is co-extensive with that of the other, but this is not always the case, and wide variations may be observed. Cutaneous analgesia is generally combined with a certain amount of deep analgesia (loss of sensibility to painful pressure), although their degrees may be unequal. Head and Thompson record a case in which an area of complete cutaneous analgesia was co-existent with deep hyperalgesia in the neighborhood of a painful arthropathy of the shoulder. "The lightest pressure stimuli caused pain, and the extent of the hyperalgesia could be marked out on the skin. Yet the superficial layers of the skin were analgesic to the prick of a pin when raised from the subcutaneous structures."

(c) *Distribution of Dissociative Anesthesia*.—Thermo-anesthesia and analgesia are usually found first in the upper extremities and thorax, and spread thence all over the body. In rare instances they commence in the lower extremities or on the face. Their distribution is nearly always asymmetrical. The borders of the cutaneous loss are not sharp, but shaded off and correspond to the limits of spinal root areas. At the same time charts sometimes show regions of dissociative anesthesia which correspond laterally to one or more root areas, but do not cover their longitudinal extent. For instance, the sensory loss in one hand may be limited above by a line encircling the forearm, so as to give it the appearance of a glove distribution. Similarly on the face a central area, including the nose, mouth, and eyes, may preserve its sensibility intact, while the surrounding regions are completely insensitive to painful and thermal stimuli. The explanation of these limitations is somewhat difficult; the presence of a cavity running through several segments and interfering, in the gray matter of each, with corresponding systems of afferent fibres connected with one posterior root after another, may throw some light on the matter.

(d) *Tactile Anesthesia*.—Sensibility to tactile stimuli and with it the power of localizing touches and distinguishing between the head and point of a pin is usually preserved until the later stages, when it is lost over areas of varying distribution. It is lost earlier, however, in that class of cases which has already been referred to as the spinal tumor class, in which no dissociation of anesthesia may be present.

(e) *Loss of Other Afferent Impulses*.—The sense of passive position and movement is often preserved until late, but may disappear progressively from one joint after another in either the upper or lower extremities. According to the observations of Head and Thompson, the power of discriminating between the two points of a compass tends to disappear *pari passu* with the sense of passive movement. Other afferent but non-sensory impulses, for instance those destined for the cerebellum, may be interfered with and defective equilibrium be the result.

(f) *Subjective Sensation*.—Subjective sensations occasionally form the initial evidence of disease, and may be of a thermal, painful, or tactile character. Lancinating pains have been described, and cramps of

considerable severity are not unknown. More curious is the subjective sensation of drenching sweat in a part which is dry. A girdle sensation occurs in this disease, as it does in many other affections of the spinal cord.

**Vasomotor and Secretory Disturbances.**—These form a very large group of symptoms, many of which, although common in syringomyelia, are not characteristic. The appearance of an affected limb may vary from time to time, now dry and pale, then moist and hyperemic. Vasomotor paralysis has the effect of making affected parts react in an exaggerated way to the temperature of the environment, with the result that the skin is overwarm and moist in the summer, and overcold, perhaps cyanotic, and dry in the winter. A cutaneous tache is of common occurrence in analgesic areas, a pin prick producing a red spot with a central white area which may last for hours, or even days, and may resemble a parasitic bite. Urticarial rashes are very prone to make their appearance with little provocation. A vasomotor disturbance of the subcutaneous tissues is largely responsible for the "main succulente" first described by Marinesco. In this the whole of the dorsum of the hand and all the fingers may swell to an extent which renders their detailed anatomy unrecognizable, while the overlying skin is tense, glossy, and smooth. Pressure produces no marked pitting. Hyperidrosis is another common feature, and often affects the analgesic areas. It may be spontaneous or easily elicited by the warmth of a hot bath. In this way half the head and neck, together with the corresponding arm and side of thorax, may be bathed in sweat while all the rest of the body is dry. Excessive lacrymation or salivation is sometimes observed in bulbar cases, and Guthrie has reported a unique instance of unilateral seborrhœa.

**Ocular Sympathetic Paralysis.**—Owing to the frequent involvement of the first dorsal and eighth cervical segments by the gliomatous process, oculopupillary symptoms occur in about 85 per cent. of cases. As a rule, the paralysis of the sympathetic fibres is unilateral, or is more advanced on one side than on the other. On the affected side the pupil is smaller and the palpebral fissure narrower than on the other side. Owing to paralysis of Müller's unstriated muscle, the eyeball is often slightly retracted. The visible effect is one of false ptosis, which disappears at once when the patient looks upward. It is stated that paralysis of unstriated muscle fibres in both the upper and lower eyelids allows of their approximation. Further examination would show that the pupil does not dilate or dilates sluggishly when cocaine is applied or when the eye is shaded, and that no enlargement takes place when the skin of the neck on the same side is stimulated.

**Trophic Disturbances.**—(a) *Skin.*—The glossy skin, or "peau lisse," represents the common atrophic variety of the cutaneous affections. Hypertrophy or thickening of the skin occasionally takes place, and this is particularly liable to lead to fissures, which deepen and ultimately ulcerate. All forms of ulceration tend to spread and perforate deeper subcutaneous structures, necrosis of bones eventually being produced. Healing is slow and sometimes impossible. In extreme degrees the

panaris of Morvan's type of the disease is the result. Keloid has been known to develop in the cicatrices of these trophic ulcers.

Among many forms of *dermatitis* which have been described, the eruption of vesicles or bullæ is one of the most common. These may appear suddenly in the night, accompanied sometimes by fever, and are distributed generally over the arms or thorax. The superficial layers of the rete mucosum are only involved at first, but when the fluid has escaped deeper ulceration generally ensues. True pemphigus is a rare but occasional complication.

Trivial injuries may lead to serious septic processes, owing to the analgesia preventing their early discovery and attention. The nails are generally thickened, striated, and brittle; sometimes they tend to drop off. Cases are on record in which nails grew in unwonted positions, while the natural nailbeds remained inactive. The hair may be altered in quality, becoming brittle and coarse, or it may disappear from large tracts of skin. A complete alopecia has been seen by the writer. The growth of hairs in unaccustomed sites has been noted.

(b) *Bones*.—In addition to suffering in the general tissue necrosis, the bones, particularly of the extremities, are liable to undergo trophic changes in the form of rarefaction or increased density. Osseous excrescences or exostoses, symmetrical in distribution, have been observed, and in at least one case suppuration has taken place in the newly formed tissue. Fragility of the long bones has given rise to spontaneous fracture, the occurrence of which is more common in the radius and ulna than in all the other bones of the body put together. General enlargement of parts of the skeleton occasionally produces a semblance of acromegaly, although it is often symmetrical and may not be associated with increase of the softer tissues. Chiromegaly is the most common example of this condition. The deformity described under the name of "thorax en bateau" by Marie and Astié is the result of osseous changes in the upper part of the sternum and upper ribs. In this there is a depression of these parts of the thoracic skeleton, which is associated with a forward and upward projection of the shoulders. It is not always combined with vertebral deformities, and therefore not dependent upon the latter.

(c) *Joints*.—Arthropathies occur in 20 to 25 per cent., the incidence on particular joints being very different to that which characterizes *tabes dorsalis*. Thus, 80 per cent. of the arthropathies of syringomyelia are found in the upper extremities, and the most frequent joint to suffer is the shoulder. As a general rule, the joint troubles are unilateral, but occasionally bilateral and symmetrical arthropathies are observed. According to Schlesinger, males present this symptom more commonly than females in the proportion of two to one. While trauma has an undoubted influence in certain instances, it is equally certain that they may appear spontaneously, and probably do so in the majority of cases.

The development of a Charcot's joint is often sudden, although it may be preceded by pains or by unpleasant sensations in the part about to be affected. The first evidence in some cases is an extensive fluid swelling involving the joint and surrounding tissues, in others a creaking or crepitus evoked by movement. The fluid is apt to disappear in the

course of a few days or weeks, leaving a relaxed and abnormally mobile joint. Fresh effusions may occur spontaneously or as a result of some slight injury, every such occurrence producing increased disorganization. The changes are of an atrophic or hypertrophic nature. Sometimes the two varieties are combined. The hypertrophic changes include thickening of the synovial membrane, ligaments, and capsule, with the appearance of osteophytes and cartilaginous masses often embedded in the softer tissues. Bony excrescences originating in the muscular insertions add to the deformities. Ankylosis has occasionally occurred. In the atrophic form the articular cartilage gradually disappears and the subjacent bone may be rarefied and destroyed so as to cause considerable shortening.

In the majority of instances these arthropathies run a painless course, but this is not invariably so in their early stages when deep analgesia may not have developed. Suppuration occurs in a certain number, and this leads to increased disintegration and perhaps to a general septicemia. Operative measures may often be carried out without an anesthetic, although sinuses very difficult to close generally persist.

*Disturbances of Control over Excretory and Sexual Organs.*—In the common type of cervicodorsal syringomyelia the interference with the control over the bladder and rectum varies with the degree of spasticity in the lower extremities. In the early stages there may be no abnormality. With moderate degrees of spasticity it is usual to find precipitate or hesitating micturition and rather obstinate constipation associated with precipitate defecation after aperient medicine. In later stages retention and incontinence are experienced. In the more rare lumbosacral form of the disease sphincter trouble may be prominent from the beginning, and in this condition the sexual power is not preserved so long as it is in the more common type. In the spinal tumor type the symptoms of a complete transverse lesion may include priapism.

**Diagnosis.**—Syringomyelia is one of the easiest and one of the most difficult diseases to diagnose. The majority of cases cannot escape recognition if a systematic examination of the nervous system is carried out. The minority may successfully simulate a number of other conditions for a time, and may even elude discovery until post mortem.

The ordinary atrophic case will seldom be mistaken for progressive muscular atrophy if the patient's sensibility is carefully tested. Vague sensory disturbances with spastic palsies, nystagmus, and sphincter troubles may sometimes suggest disseminated sclerosis, but the absence of the remitting history of the latter and the presence of trophic disorders and perhaps of congenital anomalies point to syringomyelia. Spinal hemorrhage or hematomyelia may reproduce the symptoms and physical signs of syringomyelia exactly, but the history of the acute onset will make the diagnosis. At the same time there are cases in which the evidence points to hemorrhage occurring in a syringal cavity, and thus exciting a clinical condition which was merely latent, or others in which the track of an old hemorrhage may possibly be the starting point of a gliomatosis.

By far the most difficult problems are presented by intramedullary



tumors, on the one hand, and syringomyelia with localized neoplasms on the other. The signs of a cavity extending above the level of the transverse lesion are often vague and sometimes absent, while the extension downward is generally entirely obscured by the paraplegia. Fortunately the distinction between these two maladies, equally incurable, is not a matter of vital importance. Extramedullary tumors less often cause confusion, but in any case of doubt it is better to explore than to allow any possibility of a removable growth being left.

**Prognosis.**—There is no cure, and therefore the prognosis is never favorable. The chances of life are variable, and can only be estimated by an extensive observation of the rate at which the disease is progressing. The lower the disease the shorter the life is an axiom of some veracity. Fortunately the statement can be truthfully made to patients and their friends that it is not unreasonable to expect an arrest in the morbid process, and to hope that such an arrest may occur before the paralysis has reached the stage of complete disablement.

**Treatment.**—Drugs have no power to arrest the course, and surgery cannot help us. The alleviation of pain, if there is any, and careful protection against all the septic processes to which these patients are so exposed comprise the chief treatment. In the earlier stages massage may temporarily increase the power of enfeebled muscles, but neither this nor electricity will do more. Antisyphilitic remedies should be tried when a history of lues is obtained. Belladonna and ergot will be found useful in relieving the early sphincter trouble and in diminishing painful spasms. Suppuration of bones and other tissues must be treated in the usual manner, amputation being resorted to with hesitation.

Speaking generally the patient should be advised to lead a quiet healthy life, paying particular attention to the avoidance of all injuries, however trivial they may appear to be.

### MYELITIS.<sup>1</sup>

**Definition.**—A non-systemic, diffuse, or focal affection of the spinal cord, of heterogeneous yet usually infective and extrinsic origin, disconcerting in the variety of its pathological processes, but characterized by a clinical picture, which depends for its details in individual cases more upon the segmental level than upon the exact nature of the underlying lesion.

The vascular element in the production of spinal softening has gained well-deserved recognition in recent years, and as a result of this and the prevailing view that myelitis should only be applied to an inflammatory condition, the desire to introduce such names as spinal thrombosis and myelomalacia has found expression. Syphilis now takes such an important place in the etiology of so-called myelitis that controversy has arisen as to how far the preservation of the name in its wide sense is justifiable.

<sup>1</sup> Myelitis from a true etymological standpoint only signifies disease of the spinal marrow. The suffix has, however, for a number of years been regarded as denoting inflammation. Consequently, by analogy, myelitis is generally held to mean inflammation of the spinal cord, although it has been used with a great deal of license.

The difficulties in the way of a general agreement on this and other points may be discussed here because they are of fundamental importance in arriving at a true conception of the etiology and pathology of acute myelitis. In the first place, the result of an acute or subacute focal lesion of the spinal cord is a clinical picture, which varies with the site rather than with the nature of the morbid process. It will be much the same whether the latter is of inflammatory or vascular origin, and in many cases a discrimination between the two is only possible after death, and not always then if the disease is of long standing. For this reason the term "myelitis" is convenient from a clinical point of view, and has by long usage become the natural designation for cases of the kind.

In the second place, thrombosis can never be regarded as the primary and sole factor in any of these cases. As a general rule, it occurs either as the result of disease of the bloodvessels or as one of the phases of an inflammatory process. In the latter case the primary factor is of such a character as to justify the term myelitis. In the former the disease of the bloodvessels may be either degenerative, as in certain forms of arteriosclerosis, or inflammatory, as in syphilitic arteritis.

Syphilitic thrombosis has some claim to be called inflammatory, and the clinical analysis of the conditions influencing the production of an acute paraplegia must be uncommonly close in many instances if the observer is to give a truly scientific diagnosis. Further difficulty arises from the fact that in syphilitic cases it is often impossible to differentiate clinically a spinal thrombosis from an intramedullary gumma.

The issue must also be considered from its histological side. Is it possible to distinguish, even in recent cases, a purely thrombotic from an "inflammatothrombotic" lesion? Singer, who has strongly upheld the importance of vascular disease in the genesis of myelitis, makes the presence or absence of a cellular infiltration the chief criterion upon which this question must be decided. He maintains that in cases of syphilitic thrombosis, although the larger vessels may show the usual perivascular small-celled infiltration, yet the necrosed areas are free from cellular infiltration and neuroglial proliferation. In conditions of simple inflammation, on the contrary, the excess of cells both in the perivascular lymph spaces and in the surrounding tissues is an essential and striking feature. The former condition was seen in the cases Singer described, and is undoubtedly present in many others. On the other hand, there are instances of acute spinal syphilis in which thrombosis and consequent necrosis play a very insignificant part, and in which gummatous infiltration of the tissues is so marked that many aspects of a true inflammation present themselves, with the result that the luetic origin of the lesion can only be gauged from the specific changes in the walls of the bloodvessels. Under these circumstances inflammation is at least as prominent as thrombosis in the microscopic field. If, moreover, the opportunity of examining the cord does not take place until a long time after the onset, it is more than probable that an accurate discrimination between its vascular or inflammatory origin will be impossible. Necrotic, sclerotic, or rarefied areas might be the result of either process, and a thickened, perhaps hyaline condition of the bloodvessels

is as likely in the one as in the other. Nor do the points of similarity end here. In some cases of inflammation of bacterial origin areas of necrosis without cellular infiltration, and not dependent upon vascular occlusion, are met with, and are generally supposed to be due to toxic influences.

In spite of the difficulties, there can be no doubt that at the present day *thrombosis* due to syphilitic arteritis, and occasionally phlebitis, is responsible for the symptoms of a large number of examples of acute myelitis. It is equally certain that the same criticism can be applied to cerebral syphilitic thrombosis, and it is to be regretted that the close analogy of the two conditions, differing chiefly in their site, is not preserved in their terminology.

Another point of interest now claims attention. It might be supposed that no difficulty would arise in comprising under the term myelitis all cases of spinal disease which are presumably due to the action of bacteria or their toxins, but this is not universally admitted. There are on record instances of paraplegia in which the clinical picture has suggested an acute or subacute inflammatory myelitis, but in which postmortem examination has revealed no interstitial changes, no cellular infiltration of the vessel walls, perivascular spaces, or surrounding tissues, but only degenerative changes in the more highly specialized ganglion cells and nerve fibres. The question as to how far toxic retrograde changes of the parenchymatous elements can be properly regarded as constituting evidence of myelitis is one which has raised considerable controversy. The two characteristics of this type of case are (1) oedema of the tissues and (2) degenerative changes in the nervous elements. Both may be present in the more truly inflammatory and infiltrative forms of myelitis, but they may be found in certain cases as the only changes. Storch and v. Kahlden consider that inflammatory oedema is sufficient alone to justify the use of the term "myelitis." Mayer, on the other hand, demands an acute infiltration of the vascular adventitia and perivascular lymph spaces with small round cells.

It has long been the custom to describe as "compression myelitis" the altered condition of the spinal marrow which has resulted from the pressure exerted by tumors, thickened meninges, or displaced bone. In the absence of secondary infection by bacteria it is probably more correct to look upon these changes as due to evascularization and dependent upon the ischemia produced by slow vascular occlusion. The introduction of some term such as compression evascularization of the spinal cord is not, however, likely to be welcomed. The only course is to retain the word myelitis in its broader and, etymologically, more accurate sense, preserving distinctive characters by the use of qualifying adjectives.

Acute or subacute myelitis will be considered under the following heads: (a) Acute infective (or infiltrative) myelitis; (b) acute toxic (or degenerative) myelitis; and (c) acute syphilitic myelitis.

(a) **Infective (or Infiltrative) Myelitis.**—**Etiology and Pathogenesis.**—Infective myelitis is now, at any rate, a rare affection which has no definite relation to age or sex, although young adults are probably its most common victims. It does not occur in epidemic form, nor has it seasonal

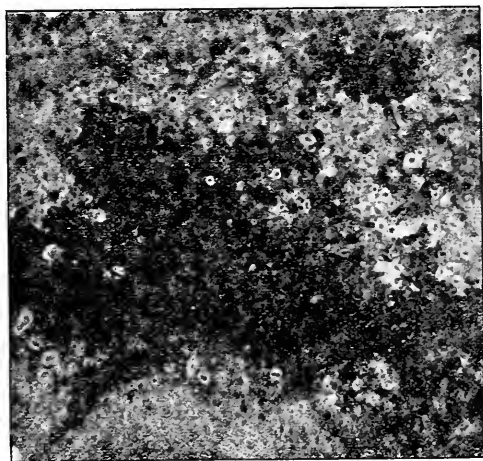
or climatic proclivities. No real importance can be given to the influence of chill, trauma, strain, or sexual excess, although it is possible that

FIG. 16



Acute infective myelitis. Vessels engorged with blood and surrounded by cells in the white matter of the cord.

FIG. 17



Acute infective myelitis. Hemorrhage into the white matter.

these may be of some predisposing or determining value in certain cases. On the other hand, the onset has not infrequently occurred in the course

of one or other of the specific infections. Barlow and Primangeli have recorded fatal cases during or following an attack of measles. Auché and Hobbs and Marinesco and Oetlinger have described its association with variola. Lepiné and Schiff have each seen myelitis occurring as a fatal complication of typhoid fever. Gonorrhœa has also been reported in a similar connection by Leyden, Gowers, and others.

The incidence of myelitis upon pregnancy or the puerperium has been of sufficient frequency to attract attention, but in many such cases the description is not detailed enough to enable one to judge whether they shall be included under the inflammatory or the syphilitic groups. There is probably a special type of myelitis, which occurs in connection with childbearing, and which will be referred to among the "toxic" varieties. It may be presumed that the disease can arise in one of several ways:

1. As an extension from neighboring inflammatory processes, particularly when the latter involve the meninges or vertebral column. A tuberculous form of myelitis occurs in connection with spinal caries and less frequently with tuberculous meningitis. Some degree of myelitis is often present in meningitis due to pneumococci, staphylococci, streptococci, and other organisms, but in these the symptoms of myelitis are largely overshadowed by those originating from the meningeal affection. On the other hand, infective myelitis practically never exists without some degree of meningitis caused by the same agent. It must then be regarded as an essential part of the disease.

2. As the result of an infection of the spinal tissues through the blood stream. This may take place either as an incident in a general pyemia or as a purely local phenomenon.

3. As the result of an infection through the lymphatics. The possibility of this is based more on the analogy of experimental pathology than on verified cases of human myelitis. The investigations of Homen and others prove that infection of the spinal cord may take place along the course of the lymphatics accompanying the spinal nerves, and that this may give rise to foci of intramedullary inflammation similar in all respects to those seen in the human cord. The paucity of clinical evidence on this point would be remarkable were it not for the difficulty attending their elucidation, but Marinesco has succeeded in tracing an infection of the spinal marrow by way of the neural lymphatics in a patient suffering from crural gangrene. Further investigation of such sources is urgently needed, and might throw light on the pathology of spinal disease following cystitis and other septic conditions of the pelvic or abdominal organs.

The bacteriology of myelitis is only in its infant stage, and is hampered by the fact that many organisms appear to be very short-lived in the tissues of the spinal cord. Streptococci and staphylococci have been found in some cases, and a diplococcus was certainly the causal agent in a case reported by T. Buzzard and Risien Russell. Marinesco has shown experimentally that myelitis can be produced by several of the ordinary pyogenic organisms.

**Pathology.**—The gross changes are generally striking and essentially the same in character, whether the extent of the lesion justifies the use of the terms, transverse, diffuse, or disseminated, in order to further

describe it. A transverse myelitis is one which is limited in the longitudinal direction to one, two, three, or, at most, four segments, and which affects the transverse area of the cord within these limits more or less completely. A diffuse myelitis involves a considerable length of the spinal marrow without definite interruption, and is often the anatomical equivalent of a clinical "ascending myelitis." The term "disseminated" implies the presence of two or more foci of the disease separated by comparatively healthy intervening tissue.

The morbid appearances vary somewhat with the length of time which has elapsed since the onset. In early cases the affected area is softer than normal, sometimes diffuent, swollen, and œdematous and generally hyperemic throughout. Hemorrhages may give a dark red or brown color to the cut surface. The definition between the white and the gray substance is frequently lost. The soft meninges on the surface of the altered parts present dilated bloodvessels, and in some cases contain serous or even purulent exudation. The exudation, if present, is usually more profuse on the posterior than on the anterior aspects of the cord. In the later cases the softening is at least as marked, but is associated with shrinkage rather than swelling of the diseased portions. Hyperemia is less evident, both superficially and on section, and the intramedullary tissue may be mottled with brown or yellowish patches. At levels distant from the seat of inflammation degenerative changes may possibly be detected in the ascending and descending tracts of the white column. In very severe lesions a segment or two of the cord may be reduced to a purulent fluid contained within the pia arachnoid membranes.

At a still later period the consistence of the diseased area is more firm, even sclerotic, and a transverse section may reveal the presence of one or more cysts. The tissues are devascularized, in part opaque white, and in part gray or translucent in appearance. The secondary degenerations in the white matter are now very evident to the naked eye.

**Histology.**—The features belong partly to the vascular tissues, partly to the neuroglia, and partly to the true nervous elements. In individual cases the relative importance may vary, but in all examples of this particular form of myelitis the vascular phenomena play some part. The two striking things are the dilatation of the bloodvessels and the excess of nuclear elements in their walls and the surrounding tissues.

**Vascular Changes.**—There is general engorgement of all bloodvessels both of large and small caliber. The adventitial sheaths and the perivascular lymph spaces show a large excess of nucleated cells. Examination of the latter under a high power proves them to be chiefly of the lymphoid type. The majority resemble the lymphocytes of the blood, but plasma cells, mast cells, and polymorphonuclear leukocytes are also present. The proportion of the latter to the total number of cells varies very considerably, and depends upon factors which are still obscure. If the lesion is of some days' duration, large cells filled with the granular products of disintegration may also be seen in the perivascular lymph spaces.

The *neuroglia* usually presents marked alterations. It is the seat of an œdema, in the early stages, at any rate, and microscopic evidence of this is either negative or positive. In the former case empty spaces or

lacunæ represent the fluid which has been absorbed in the process of hardening; in the latter these spaces are filled with an amorphous granular material deposited from the albuminous fluid by which the tissues have been bathed. The neuroglial cells undergo great changes. Many swell to a large size and display two or more nuclei which may be vesicular. The cell plasma is liable to vacuolation, and the processes may be thickened or lost. In addition to variety in size and shape, the neuroglial cells are greatly increased in number, especially in the neighborhood of the dilated vessels and their lymphatics. Among them are also many migratory cells from the blood, which are not always easily distinguished from those of neuroglial origin. Hemorrhages into the tissues and masses of granular debris are also common in these situations.

*Changes in the Nervous Elements.*—The nerve cells of the gray matter undergo changes which may be partly due to the œdema, but which are mainly the result of toxic influences. They become swollen, rounded, and homogeneous in appearance. The chromatin granules disappear and the nuclei lie excentrically or protrude from the cell periphery. The axis cylinder processes are swollen and often broken. The earliest changes in the white matter are those which affect the myelin of the nerve fibres. This swells and rapidly loses its power of staining deeply by the Weigert-Pal method. The Marchi method reveals the presence of fatty changes. The axis cylinders are also enlarged in transverse and irregular in longitudinal outline. In sections the myelin is apt to disappear in places, and the axis cylinders remain naked and unsupported.

Three weeks after the onset a Marchi preparation shows that a great deal of fatty degeneration has taken place and that much of the fat has been taken up by large granular cells, which are probably of neuroglial origin. Sections from other parts of the cord show in a striking manner the ascending and descending degenerations in the white columns.

At a later period these degenerations are visible only when the Weigert-Pal method is used, and by this time the acute changes in diseased areas have been replaced by those which represent attempted repair. The vessels have lost much of the perivascular cell infiltration, but their walls are thickened and perhaps hyaline in appearance. Proliferated neuroglial tissue has replaced the nervous elements which have been destroyed, and possibly cysts may indicate areas of necrosis. In the sclerotic zones the number and distribution of vessels may suggest that there has been some new formation of the latter. Secondary degeneration appears in the anterior spinal roots, the efferent fibres of the peripheral nerves, and in the muscles innervated by those segments of the cord in which the gray matter has been seriously involved.

**Symptoms.**—These are dependent on the site and extent of the spinal inflammation, and vary accordingly. In the majority the onset is associated with some constitutional disturbance, pyrexia, anorexia, and perhaps vomiting. The more severe paralytic troubles are usually preceded by pain in the back of a burning character and by paresthesia in the limbs or in those parts about to be the seat of serious disorders of motion and sensation.

*Transverse.*—A focal lesion affecting more or less completely the whole transverse area of the cord produces a clinical picture which varies according to whether the cervical, dorsal, or lumbosacral regions are involved.

(a) *Cervical.*—A focal myelitis high up in the cervical region is rare, and usually rapidly fatal, owing to paralysis of the entire respiratory musculature. More common are cases of myelitis affecting the cervical enlargement. In such a case there may be atrophic flaccid paralysis of the hand muscles and the flexors of the wrist and fingers, while the extensors of the wrist and the muscles moving the elbows and shoulders remain intact; at the same time there is partial or complete spastic paralysis of all muscles below the arms, including those of the trunk and legs with the exception of the diaphragm. Such a distribution of paralysis indicates a lesion of the eighth cervical and first dorsal segments, and is associated with diminution or loss of cutaneous sensibility in the area supplied by those segments and by all the segments below that level.

Respiration is embarrassed on account of interference with the intercostal muscles, and control over the sphincters is impaired. Absence of the abdominal reflexes, exaggeration of the tendon jerks in the lower limbs, clonus, and extensor responses are also observed. The incidence of the lesion on the eighth cervical and first dorsal segments would probably cause irritation or paralysis of the oculopupillary fibres and lead to widening or narrowing of the pupils and palpebral fissures.

The above picture indicates a partial lesion. If the morbid process produces a complete discontinuity of function, the paralysis below the lesion is of flaccid type, and accompanied by loss of all tendon jerks, at any rate for a considerable time. Moreover, the anesthesia is complete for all forms of sensation, and trophic changes in the skin, especially bedsores, are readily excited. Total paralytic incontinence of urine and feces adds to the difficulty in preserving an intact skin. Splanchnic paralysis giving rise to distension of the hollow abdominal viscera is another result of any transverse lesion above the dorsal region.

(b) *Dorsal.*—In this case the arms escape, and it is generally easier to locate the upper limit of the lesion from the sensory than from the motor disturbances. Paralysis of the lower half of the rectus abdominis may, however, give rise to drawing up of the umbilicus when the patient attempts to sit up. Examination would then reveal the presence of the epigastric and the absence of the hypogastric superficial reflexes, which, with the other observation, points to the ninth dorsal segment as the highest one involved. The atrophic paralysis due to destruction of the lower motor neuron elements is not so easy to detect in the trunk muscles as in those of the limbs. The paraplegia resembles that of the cervical cases; it is spastic with partial and flaccid with complete lesions.

If the inflammation is limited to either the left or the right half of the transverse area of the cord, the signs are more or less those which have already been described under the name of Brown-Séquard's paralysis. It is, however, rare for an inflammatory lesion to be so limited as to reproduce exactly the symptom complex of an experimental hemisection of the cord.



(c) *Lumbosacral*.—Myelitis of the lower parts of the cord may give rise either to a mixed spastic and atrophic or to a purely atrophic type of paraplegia. Rapid paresis, atrophy, and reaction of degeneration in groups of muscles associated with areas of impaired sensibility, loss of tendon reflexes, and paralytic incontinence of the sphincters are the characteristic features. If the disease is limited to the upper part of the enlargement, atrophic palsy of certain muscles of the hip and thigh may be combined with spastic paresis of those moving the ankles and toes. In such a case the knee-jerks may be absent, and yet ankle clonus and extensor plantar responses be easily elicited. A clinical picture of this kind can be readily understood if the segmental innervation of the musculature is remembered.

The conditions so far described represent the fully established disease in its early days. A certain number of cases run a rapid course to a fatal termination, others last a few weeks, but succumb finally to some complication. In those which survive, recovery of function modifies the clinical picture in varying degree. When the recovery is insufficient to allow of walking or standing, the resulting condition, in the cervical and dorsal cases, is one of spastic paraplegia in a bedridden patient who often suffers from painful flexor spasms of the lower extremities, from a sense of constriction at a level varying with the site of the lesion, and from retention or incontinence of urine and feces. Improvement in the cutaneous sensibility usually precedes and remains in advance of any improvement in voluntary power. Such a patient may live for many years and enjoy good general health if surrounding circumstances are favorable. In less severe instances a remarkable return of function may be witnessed and the patient may recover with a normal gait or more often with some degree of spastic paraparesis, necessitating, perhaps, the aid of a stick. Control over the sphincters may be entirely regained, or any deficiency in this respect may only be noticeable in a tendency to precipitate micturition or defecation after aperients, or after drinking much fluid. Sexual impotence is the rule in most cases of myelitis. Complete restoration of sensibility is general in such cases, and the last evidence of the disease to disappear is the extensor type of plantar response. In the lumbosacral type the prospect of complete regeneration of the atrophied muscles is not bright and the more serious interference with the sphincters adds to the gravity. Considerable degrees of recovery, resulting in a condition of partial disablement not unlike that which follows lumbar poliomyelitis, may be expected in patients who survive the early stages.

*Disseminated Myelitis*.—This must be regarded more as a multiple of the transverse type than as a separate disease. It is very rare, and can only be recognized clinically when, for instance, a focal lesion in the dorsal region of the cord causing a spastic paraplegia is associated with an atrophic paralysis of one arm or with a patch of anesthesia above the level of that which is the result of the dorsal myelitis. Further evidence may be afforded by the appearance of some affection of one or other of the cranial nerves.

More often dissemination of the patches of inflammation is only

revealed post mortem, the clinical picture having suggested a transverse myelitis corresponding to the highest focus in the spinal cord. In other cases the clinical course was that of an acute ascending myelitis, although examination of the spinal cord showed considerable dissemination of the lesions. Possibly there is no room for the two separate terms, disseminated and ascending, the one referring rather to the anatomical and the other to the clinical aspect of the same kind of case.

*Acute Ascending Myelitis.*—This forms a morbid entity which can be readily recognized during life. It is characterized by a progressive paralysis of motion and sensation beginning in the lower extremities and more or less rapidly climbing up the body, segment by segment, until involvement of the respiratory musculature threatens life, and not infrequently produces a fatal termination. The extension of the disease to the brain-stem is occasionally indicated by the implication of cranial nerves before death, and the occurrence of optic neuritis has been recorded in non-fatal cases when all other symptoms have been referable to the affection of the spinal cord. With the more usual employment of lumbar puncture the bacteriology is likely to become better known.

(b) **Toxic (or Degenerative) Myelitis.**—It has already been stated that infective or infiltrative myelitis is by no means a common disease. Toxic myelitis must be very rare, and the name can only be applied to a small proportion of all cases. Very little is known about its origin, although it appears from experiments that bacteria and their toxins are able to produce similar conditions in the lower animals. Moreover, it seems possible that the same organisms may at one time or under one set of circumstances produce an infiltrative and at another time or in other conditions determine a toxic form of myelitis. It is equally certain that the effect of some organisms upon the spinal cord is in part to excite inflammation in the interstitial tissues, in part to produce retrograde changes in the nervous elements.

Speaking generally, toxic myelitis differs from infective myelitis in the less acute, more subacute onset and course, the small amount of constitutional disturbance, and perhaps the power of more complete recovery. This last point must necessarily be one rather of speculation than of actual fact, but it is natural to suppose that toxic changes, which are known to be capable of recovery, leave less permanent effects than inflammatory lesions, resulting as they usually do, in necrosis and sclerosis. Of the etiology, little can be said, because, as a rule, the toxic nature of the process is not discovered until after death. As an exception must be mentioned the cases of toxic myelitis associated with pregnancy. The close association is shown by the improvement in the myelitis at the birth of the child and by its recurrence in subsequent pregnancies.

Probably some inorganic substances, lead, alcohol, and certain poisonous gases, are capable of producing a toxic myelitis, but if so they are not commonly concerned in this connection, and the part they play is referred to elsewhere. Diphtheritic toxins are among the possible causal agents, but it is doubtful whether they ever excite a general toxic myelitis. Among the cases of unknown bacterial origin must be grouped those which for many years have gone by the name of Landry's paralysis.

**Pathology.**—Gross changes are not very obvious, but some œdema of the spinal tissues with consequent softening may be observed. On cross-section some degeneration may be seen in the white columns. Microscopically, the chief changes consist in swelling of the myelin sheaths and of the axis cylinder processes, especially in the white matter. The change is generally patchy and shows no tendency to follow a systemic distribution. In addition there are toxic changes in the nerve cells as well as in those of the neuroglia. The ganglion cells swell, become rounded and chromatolytic with excentric nuclei. The neuroglial cells also become round and enlarged, but do not show the great increase in number characteristic of the infective form. In the early stages the Marchi stain demonstrates fatty changes in the myelin sheath; later on the disappearance of groups of these sheaths and the dilated spaces they leave give a somewhat “worm-eaten” appearance to the affected areas.

**Symptoms.**—These depend upon the site of the disease and resemble those of the infective type in all respects. The onset and course are generally less acute, and the power of recovery is probably greater, but in the majority of cases it is quite impossible to differentiate the two conditions at the bedside. Motion, sensation, and sphincter control are all impaired and the relative amount of muscular spasticity and atrophy varies with the degree in which the upper and lower neurons are involved in the morbid process. In a case described by Rhein the clinical picture resembled that of a subacute ascending myelitis of the inflammatory kind. Post mortem, only cell changes and myelin degeneration were found to account for the symptoms. The cases in pregnant women are also characterized by an ascending progress of their symptoms. In fatal instances bulbar symptoms followed those of spinal origin.

(c) **Syphilitic Myelitis.**—**Etiology.**—Syphilis is by far the most important factor in the production of myelitis, and for this reason cases of syphilitic myelitis easily outnumber those of other origin. Singer found that out of 19 cases of myelitis admitted in the course of two years into the National Hospital (London), 15 had a definite history of syphilis. In 12 of them the onset of myelitis occurred within three years of the primary infection. In the opinion of the writer these figures, roughly 80 per cent., do not exaggerate the preponderating causal influence of syphilis. *Males* are much more frequently affected than females, and the greatest susceptibility prevails during the third, fourth, and fifth decades. It is probable that *age* has also some influence upon the rapidity with which myelitis follows the infection, the interval being shorter in the younger patients. This interval varies from a few months to many years, but the liability to the disease diminishes after three years have elapsed. Gummatous meningomyelitis may also be the result, although rarely, of congenital syphilis. In at least 75 per cent. of cases of syphilitic myelitis the incidence of the disease is upon the dorsal region of the cord and more particularly upon the middle third of that region. This may be explained by the fact that this area is more poorly supplied with large contributing root vessels than any other, and is in distinct contrast to the incidence of the infective type which favors the lumbar and cervical enlargements at least as frequently as the dorsal region.

It may be doubted whether any other factor than that of lues is of material importance in the etiology of this form, but it seems possible that great fatigue, exposure, and perhaps alcoholism may have an exciting influence. It is certainly common among soldiers.

**Pathology.**—The influence of the syphilitic virus upon the spinal tissues is not a simple one, but is exerted in a number of different ways. Syphilitic myelitis is really the manifestation of more than one morbid process, and each variety may be represented to a greater or less degree in one case of the disease. It may be said that the spirochæte produces its results in part by its power of causing changes in the walls of bloodvessels with consequent effects upon the local circulation, in part by the diffusion of toxic substances capable of exciting profound influence upon certain of the tissues, and in part by exciting a cellular infiltration of an inflammatory or granulomatous character.

Any or all of these results may be concerned in any particular case, but, as a rule, one or other has more or less preponderating importance. It must always be remembered that morbid changes may be silently at work for a considerable time before some sudden catastrophe, such as the occlusion of an important bloodvessel, provides a suggestion of acuteness to the illness to which it is in reality hardly entitled. For much the same reason it is the rule to find evidence of syphilitic changes in the bloodvessels and meninges over a much wider area of the cord than the segment or two forming the site of the lesion which has determined a disturbance of function. Although concerned for the moment with the intramedullary changes, the fact that these are often secondary to, or intimately associated with, syphilitic processes in the surrounding tissues, especially with those in the dura mater, must not be lost sight of.

The incidence of syphilitic myelitis is much more frequent in the dorsal region than elsewhere, but wherever placed the focus of disease is generally well defined. The lesion is usually single and generally limited to a very few segments, so that it may be included under the term transverse myelitis. The diseased area is recognized by its soft consistence and possibly by some slight alteration in color. The pia arachnoid overlying it may be milky and contain distended veins, but the milkiness may not be more noticeable at the level of the softening than elsewhere. In long-standing cases there is marked narrowing in the caliber of the cord, as well as a wrinkled condition of the membranes covering it. Atrophy of anterior roots arising from the softened segments may be detected. Adhesions connecting the dura mater with the leptomeninges are of frequent occurrence.

**Histology.**—*The Bloodvessels.*—The vascular changes may be divided into at least three chief types: (1) Moderate perivascular and adventitial cell proliferation with thickening of the intima of sufficient degree to produce diminution or complete occlusion of the lumen. This may be very general throughout the cord or confined to the area of softening. The proliferation of small round cells may only be slight, and yet the walls of the vessel may be very thickened and perhaps hyaline in appearance. This would suggest that the morbid process has been subacute or chronic, and that the acute onset of symptoms has been due essentially

to a rapid ischemia originating in a local obstruction or following on a general lowering of blood pressure. (2) Excessive perivascular cellular infiltration with little or no endarteritis. In this case there may be no evidence of actual obstructive thrombosis, although slowing of the circulation and blood stasis is suggested by the excess of white corpuscles in the vessels. On the other hand, the changes in the neighboring tissues are probably due to a toxic necrobiosis or to an extension of the cellular proliferation from the immediate precincts of the vessels. (3) Paralytic vasodilatation with very moderate perivascular infiltration and with no obvious thickening of the vessels. Retardation of the blood stream with capillary thrombosis and profound alterations in the nervous elements are the characteristics of this change. The general appearance produced is one of hyperemia of the white as well as of the gray matter associated with retrograde changes in myelin sheaths and ganglion cells.

*The Neuroglia.*—This offers the greatest amount of resistance to the vascular changes. In areas of complete necrosis it succumbs with everything else, the cell nuclei rapidly lose their staining reaction, and the cells as well as fibres taking part in the general liquefaction. In other regions the neuroglial cells survive when the more specialized structures are destroyed. Under these circumstances they are enlarged, rounded, and often multinuclear. In later stages they proliferate and take a leading part in the process of repair, sending out long interlacing fibres in all directions. It is not improbable that they give origin to at any rate some of the large granular cells, which act as scavengers and which, at first distributed in the tissues, are later collected in large numbers in the perivascular lymphatics. With the neuroglial changes must also be mentioned the gummatous infiltration which occasionally goes hand in hand with the vascular phenomena. In some instances this forms the predominant feature, and a true neoplastic gumma, with the usual central degeneration and caseation, occupies a considerable part of the transverse area of the cord. In others the aspect is that of a gummatous meningomyelitis, either involving the cord in the form of a ring or penetrating its substance at one or two points.

*The Nervous Elements.*—Demyelination of the nerve fibres and retrograde changes in the ganglion cells are the results either of the circulatory disturbances or of the diffusion of toxins. The myelin sheaths swell, disintegrate, and finally disappear, leaving axis cylinders in a naked, unsupported, but often swollen and perhaps varicose condition. The ganglion cells undergo the usual changes in shape, in granulation, and in nuclear displacement, which have been described. These alterations are local and do not extend into distant parts of the cord.

*The Leptomeninges.*—Round-cell infiltration of the pia arachnoid is almost universal in the syphilitic form, and is usually associated with varying degrees of meningeal thickening. The arteries are often hypertrophied, and the veins may display an obliterative phlebitis. The walls of the latter not infrequently present a laminated appearance, layers of round cells being separated from each other by strands of connective tissue, so that their bulk is as great as that of the arteries.

The ultimate results resemble those of other forms of myelitis. The

processes of repair, of sclerosis, of new vessel formation, and perhaps of cyst or cavity production are essentially the same, and an examination of a focal lesion a year or more after the acute stage might reveal nothing which would enable one to speak with certainty as to its exact origin.

The type of syphilitic myelitis which has been referred to as transverse is the common one, and it is rare to find a diffuse myelitis of acute or subacute syphilitic origin. Occasionally a few patches of disease scattered through a few segments may invite the term "disseminated," but the clinical picture even then is rather that of a transverse myelitis. On the other hand, it is not rare for a spinal syphilitic lesion to be associated with a cerebral one of the same nature.

**Symptoms.**—Perhaps no disease has a more characteristic onset than syphilitic myelitis. A man in good health, for no reason that he can see, finds himself unable to empty his bladder, or finds one or both legs heavy and weak, numb and "prickly." He is reassured in a few hours by the disappearance of this trouble, and it may be two or three days or two or three weeks before anything further occurs. Then, perhaps after an unusually long day's work, or perhaps with no departure from the ordinary routine, he rapidly, sometimes suddenly, loses power in his legs, loses his ability to micturate, and is conscious of a numbness from his waist downward. Throughout this period he has experienced no feeling of illness, and unless he is anemic and sallow, as so many victims of recent syphilis are wont to be, has been regarded by his relations as strong and healthy. Even as he lies in his paraplegic condition his appetite and temperature are little disturbed. Such a history is common enough, but it may be varied by the absence of any premonitory symptoms or by the more gradual development of the paralysis. For instance, one leg may be paralyzed some hours or even a day or two before the other, and very occasionally the disablement is limited from the first more or less completely to one lower extremity, with the production of a modified form of Brown-Séquard's paralysis. Not uncommonly the patient has gone to bed in his usual health, and on getting up in the morning falls to the ground. Pain is not a prominent feature; it may be absent altogether, or the patient may complain of a burning sensation at a particular spot on the spinal column. A girdle sensation corresponding roughly to the upper border of paralysis may be present early or may supervene in the later stages, but paresthesia referred to the paralyzed limbs is frequently experienced. Such is the onset of a dorsal syphilitic myelitis, and the results of an examination will correspond more or less exactly to those alluded to under infective myelitis of the same region. According to whether the physiological continuity of the spinal cord is completely or incompletely severed at the site of the lesion a flaccid or a spastic paraplegia will present itself. In the former case paralysis will be complete from some level on the trunk downward, the legs will be extended, toneless, and powerless; all deep and superficial reflexes will be abolished. The bladder may at first empty itself at intervals as a reflex involuntary act without the consciousness of the patient, but later will dribble more or less continuously or whenever any change of posture is attempted or the abdomen is pressed upon. The relaxation of the anal sphincter

may be realized by the introduced finger; solid feces may be retained, but diarrhœa will lead to constant soiling. Incomplete priapism is sometimes observed. Loss of sensibility to all forms of stimuli exists up to the segmental area corresponding to the spinal disease, and a band of hyperesthesia may be present just above that level. The skin of the lower extremities is dry and sometimes œdematous. Of extreme and urgent importance is the liability to bedsores and cystitis.

In the spastic cases the lower extremities are usually flexed at the hip and knee, and strong adductor spasm brings the limbs in apposition. The tendon-jerks are lively; ankle clonus and extensor responses are easily elicited. The mere handling of the limb or the movement of bedclothes suffices to excite painful clonic spasms, which may last for some little time. The relation of the sensory disorders to the paralysis is very variable, but sensation is nearly always less affected than motion. Constipation and retention of urine with overflow incontinence are the characteristic sphincter troubles in this type.

In the more rare cervical and lumbosacral cases the symptoms are those already described under infective myelitis.

A few words only are necessary for the purpose of indicating the course. A flaccid paraplegia may become spastic in the course of a few days or a week or two. Rarely does a case of the flaccid type survive long in that condition, a fatal termination being usual. The spastic cases, under appropriate treatment, display a general tendency toward improvement and the less complete the initial paralysis the more favorable is the ultimate outlook. Only a very few cases recover completely, but a considerable number attain a condition in which they are able to get about with more or less difficulty and with greater or less degrees of spastic gait. Not infrequently the sphincter troubles prove obstinate.

**Diagnosis.**—This can be conveniently divided into two parts: (1) The diagnosis of acute myelitis from other forms of paralysis, and (2) the discrimination between the various forms of acute myelitis.

1. The diagnosis of acute myelitis from other forms of paralysis is not attended, as a rule, with much difficulty in the simple transverse cases. The rapid onset of a paraplegia with a well-defined upper limit to the motor and sensory disturbances could only result otherwise from some sudden pressure exerted by displaced bone or by a tumor. In the very exceptional cases in which symptoms of compression are really acute there is usually some indication of the gross nature of the lesion. Moreover, pain is generally a more prominent feature in these cases. Compression paraplegia developing in a patient who is suffering from cancer may suggest acute myelitis if the possibility of compression is forgotten. More deceptive are the attacks of acute, although often incomplete, paraplegia which form part of the course of disseminated sclerosis, and which sometimes constitute the earliest symptom of that disease. The absence of a well-defined upper limit to the paralysis and the presence of some unsuspected symptom or physical sign above the level of the lesion will generally clear up any doubt. The observer should pay particular attention to the cranial nerves and keep a sharp lookout for nystagmus, diplopia, or pallor of the optic disks, the presence of any

one of which would point strongly toward the diagnosis of disseminated sclerosis. An acute form of multiple sclerosis has occasionally been described, and may require careful differentiation from acute disseminated myelitis. A lumbosacral myelitis with its atrophic changes in the lower extremities may simulate an acute poliomyelitis until the presence of anesthesia and of severe sphincter troubles is recognized.

Only a few years ago the distinction between myelitis and *peripheral neuritis* was a prerogative of the expert. Nowadays it is hardly necessary to draw attention to the differences between the two diseases. In the dorsal and cervical cases no mistake can arise. In the lumbosacral form of myelitis it must be remembered that the distributions of sensory and motor phenomena follow a segmental arrangement, and have not a special incidence upon the peripheral parts of the limbs as they have in neuritis. Furthermore, the sphincter disturbances of the myelitic, and the muscular tenderness of the neuritic cases help to distinguish the two conditions. Finally, a case of peripheral neuritis is not very often sufficiently acute to give rise to the suspicion of an acute myelitis.

In all cases of myelitis lumbar puncture should be performed, and the examination of the cerebrospinal fluid will sometimes settle any doubts.

Many cases of dorsal myelitis have been regarded in their first stage as functional or hysterical, especially when young women have been the victims. Loss of power and sensibility, very brisk tendon-jerks, pseudoclonus, and even retention of urine may be found in functional paraplegia, but incontinence of urine, of feces and, above all, an extensor type of plantar reflex serve as infallible signs of an organic basis for the paralysis.

2. A diagnosis of transverse myelitis having been arrived at, the next question to be answered really amounts to this: Is it syphilitic or the result of some other infective process? The history, or absence of history, of syphilis must of course be allowed due weight, and when an undoubted luetic record exists it will necessarily indicate the line of treatment. In doubtful cases the mode of onset affords some guide. With those of syphilitic origin premonitory symptoms are often observed over a considerable period before the actual development of the disease, and the initial stage is generally associated with no constitutional disturbance. In the infective type, on the other hand, warnings are less common, and, when they do occur, precede by a few hours only the onset of paralysis. Moreover, some degree of general malaise, general pain, pyrexia, and perhaps vomiting are the rule rather than the exception in these patients. The site of the lesion is not of very great diagnostic significance, but, *ceteris paribus*, the dorsal region favors the syphilitic, and the cervical enlargement the infective form. The examination of the cerebrospinal fluid may or may not be of assistance. The presence of organisms or the discovery of the *Spirochæta pallida* would of course be more or less decisive if secondary infections could be excluded. A preponderance of polymorphonuclear leukocytes would point to a non-syphilitic virus, but a lymphocytosis does not afford definite proof of the luetic nature of the lesion. A positive Wassermann reaction in the blood associated with a positive or negative reaction in the cerebrospinal fluid



is the most satisfactory evidence of syphilis, but a negative reaction in both fluids does not necessarily exclude it.

If syphilis is suspected careful search should be made for corroborative evidence in other parts, and the pupillary light reaction in particular should be the object of investigation. A myelitis not infrequently represents one feature or phase of cerebrospinal syphilis. The writer has seen tabes, myelitis, and cerebral thrombosis present in one subject. Equally important in the infective type is the discovery of some source for the secondary invasion of the cord, and the possibility of gonorrhœa as well as of septic processes must be diligently inquired into.

The acute ascending types of myelitis, whether of infective or toxic origin, present a clinical picture, which may for a moment suggest a Landry's paralysis or an acute poliomyelitis, but the presence of sensory disturbances and of sphincter trouble suffice to exclude the latter diseases.

**Prognosis.**—The general outlook has already been indicated in describing the progress of the different forms, but certain principles may be deduced which should be of use in an individual case. The severity of the lesion is of importance both as regards the prospect of life and the hope of useful degrees of recovery. A complete loss of conducting power in the cord at the site of disease, although it may only be temporary, is a menace to life whenever it presents itself, and the higher the level of the lesion the greater the immediate danger. Interference with the respiratory musculature may lead to a rapid fatality, and even when the dorsal region is the part affected the tendency to severe decubitus and to cystitis with its renal complications constitutes a grave element. Should the patient survive these dangers, the prospect of good recovery in such severe cases is a very poor one. Instances of lumbar myelitis are particularly hopeless in regard to the possibility of regaining the power of walking, and their irremediable sphincter troubles usually determine an early death. Less complete paraplegias may be confidently expected to show marked improvement, but only after the effects of treatment, especially in the syphilitic cases, have been gauged, can any precise estimate of the ultimate result be formed. It must be stated that cases of spastic paraplegia resulting from myelitis are disappointing to themselves and to their physicians. Improvement sets in and goes on up to a certain point, and then progress is arrested and a stationary condition, obstinate to all forms of treatment, is maintained.

**Treatment.**—*Infective and Toxic Cases.*—In the acute stage the patient must be given complete rest preferably, and often imperatively, on a water bed. Changes of posture are necessary for the prevention of sores, but cannot be seriously recommended for any influence they can exert upon the morbid process. The supposition that the volume of blood in the spinal cord is lessened by placing the patient on his face, or that such a depletion, if it occurs, is beneficial, does not appear to have any reasonable basis. On the contrary, the determination of blood to the site of infection must be regarded as an essential factor in Nature's effort of resistance and the effect of gravity in altering the hyperemia as insignificant. Immediate attention should be paid to the bladder, and the urine drawn off if there is retention. Free action of the bowels

should be secured at least every alternate day after an initial purge, and the greatest care must be taken to keep the skin clean. If pyrexia and pain are present, a diaphoretic mixture containing salicylates or quinine and a light fluid diet are indicated. There is no reason to believe that the application of heat or cold to the spine can be of use, and disturbing the patient for the purpose of cupping is probably more harmful than beneficial. A lumbar puncture should be performed at the earliest opportunity and an examination of the fluid carried out. As soon as acute symptoms have passed off all paralyzed parts should be rubbed and moved every day, and each joint prevented from becoming fixed. In the dorsal cases the spastic muscles need no treatment, but in the lumbosacral cases atrophied muscles of the lower extremities should receive electrical treatment, provided that this can produce contractions. The kind of current must be chosen accordingly. A careful watch must be kept upon the urine and any sign of cystitis combated by means of hexamethylenamine by the mouth and by irrigation of the bladder with some antiseptic solution. The sphincter trouble may in the less severe cases be favorably influenced by giving belladonna internally, and the same drug combined with extract of ergot may relieve the painful flexor spasms which are particularly liable to be troublesome at night, and when sleep is much disturbed by them the use of such drugs as veronal, sulphonal, and even morphine is not only justifiable, but urgent.

As the patient gains in strength more energetic massage and passive movements may be carried out, and he should make every endeavor to perform movements on his own account. It is his duty to force impulses, as it were, through the block on the conducting lines or to find some other way round for the resumption of traffic. This side of the treatment is apt to be forgotten in the modern craze for massage and electricity. If the patient can be induced to attach less importance to the energy displayed by the rubber and to impart more energy into his own efforts at initiating voluntary movements, the medical attendant will have gained valuable coöperation. When the limit of improvement by these means appears to have been reached a change to some suitable health resort should be advised.

*Syphilitic Cases.*—Antisyphilitic treatment can in most cases be commenced as soon as paralysis sets in, or, better still, during the premonitory stage. Both mercury and iodide of potassium should be given, the method of administration being of minor importance. Beginning with 5 grains (gm. 0.3) three times a day, the iodide of potassium should be increased rapidly until 20 to 30 grains (gm. 1.3 to 2) are taken in each dose. The mercury may be given by the mouth in the form of perchloride or the red iodide, by inunction, or by hypodermic injection, but it is not advisable to apply the inunction or the injection to anesthetic parts. This should be pushed to salivation and renewed after an interval of two or three weeks. Unless there are contraindications in the way of serious cardiac or renal disease, intravenous injections of salvarsan should be given early in addition to the mercurial treatment. Two or more injections may be given at intervals of a week, according to the effect produced upon the Wassermann reaction, and the condition of

the cerebrospinal fluid. Bearing in mind the fact that the victim of syphilitic myelitis is prone to other manifestations of cerebral or spinal syphilis, and that there is no criterion as to when that liability is successfully eradicated, the patient should undergo a course of mercurial treatment two or three times a year for an indefinite period.

Further injections of salvarsan may be necessary if indications of further syphilitic lesions appear, or if the Wassermann reaction of the blood does not remain negative.

### CHRONIC MYELITIS.

There are perhaps few facts more deserving of recognition in neurological medicine than the extreme rarity of a primary chronic myelitis. One meets with patients who exhibit the results—generally a stationary spastic paraplegia—of a former acute or subacute myelitis, but rarely, if ever, does one encounter a case in which slow progressive symptoms of a spinal affection justify the diagnosis of a chronic myelitis. If such a diagnosis suggests itself every effort must be made to exclude (1) the various forms of systemic sclerosis, such as subacute combined sclerosis, amyotrophic lateral sclerosis, etc.; (2) disseminated sclerosis; (3) syringomyelia; and (4) compression paraplegia due to tumors or vertebral disease. It is to the exclusion of spinal compression that attention must especially be paid, because the life of the patient may depend on the accuracy of the opinion.

It may well be asked whether, if the chronic stages of acute myelitis and the list of diseases quoted above are put on one side, there is any condition which deserves the name of chronic myelitis. The answer to this is in the negative if an exception is made in favor of certain ill-defined and rare cases of syphilitic spinal diseases to which reference will shortly be made. In other words, slowly progressive symptoms pointing to a lesion of the spinal cord indicate in the vast majority of cases one or other of the diseases above mentioned, and not a chronic myelitis. It is true that a chronic myelitis secondary to some infective process, such as tuberculous disease of the vertebræ and spinal membranes, may be induced, but the primary cause must be firmly established before the chronic myelitis can be legitimately inferred.

In former days the term chronic myelitis was used as a cloak to ignorance, and many cases of multiple sclerosis, of subacute combined sclerosis, and particularly of compression paraplegia went under that designation. The importance of a general recognition of this is especially obvious in cases in which there is evidence of a focal transverse lesion of the spinal cord with symptoms increasing in intensity. These must not on any account be labelled chronic myelitis, but, speaking generally, must be submitted to surgical exploration in the hope that the removal of a tumor pressing upon the cord may change the prognosis of the case from that of a progressive incurable paraplegia to one which holds out good prospect of a complete or partial recovery. Emphasis having been laid upon the rarity of any spinal disease justifying the diagnosis of chronic myelitis, a condition which is known under the title of Erb's

syphilitic paraplegia may be briefly described, although the writer has some doubts as to how far a separate morbid entity answering to that name may fairly be recognized.

**Erb's Syphilitic Paraplegia.**—In 1892 Erb sought to establish a type of disease to which he gave the name "syphilitic spinal paralysis." According to his view the disease was a form of spastic paraplegia characterized (1) by its syphilitic origin; (2) by the marked exaggeration of the tendon reflexes as compared to the moderate degree of muscular rigidity; (3) by bladder trouble, which might precede by a long time the other symptoms; (4) by distinct paresthesias of subjective rather than objective character; and (5) by the gradual development of the disease and its tendency to improve under appropriate treatment. Rumpf had already shown that all these symptoms could be the result of syphilitic disease of the bloodvessels of the cord, though he had not laid so much emphasis on the activity of the reflexes in relation to muscular rigidity. At the present time it must be confessed that this point is not characteristic of Erb's type, because it may be observed in the late stages of acute myelitis and under other conditions.

The occurrence of this form of spastic paraplegia with a gradual onset and progressive course must be very rare, but it is interesting, as it provides at any rate one possible form of chronic syphilitic myelitis. It would be suggested in any particular case by a history of syphilis, and its diagnosis would be readily confirmed by a lumbar puncture, with examination of the fluid, and also by the results of antisyphilitic treatment. In a case of the kind which came under the writer's observation the bladder trouble anticipated the other spinal symptoms by many months, thus raising the suspicion that some urethral disease was responsible for the difficulties associated with micturition.

**Pathology.**—Little is known about the anatomy of these cases, because the majority, which have been called examples of Erb's syphilitic paraplegia during life, have been shown postmortem to be really the subjects of a transverse syphilitic myelitis in the dorsal region of the cord, with the usual ascending and descending tract degenerations. Other cases are the result of diffuse syphilitic disease of the spinal vessels, sclerosis taking place in areas which are poorly nourished. Finally, it is stated, without much positive evidence, that the syphilitic toxins may induce a degeneration of the pyramidal tracts and of the long ascending tracts in the posterior and lateral columns without the intervention of any specific changes in the bloodvessels or meninges of the cord.

**Diagnosis.**—This must depend on the progressive character of the symptoms and on the results of lumbar puncture and antisyphilitic treatment. The absence of pain serves to distinguish it from most, but not all, cases of compression due to tumors or vertebral disease. The definite limitation of motor and sensory disturbances to parts of the body innervated from below a certain level in the spinal cord must always suggest a focal lesion, and not a diffuse affection of the long tracts in the cord, such as is found in Erb's syphilitic paraplegia.

**Prognosis.**—Appropriate treatment tends to ameliorate the spinal symptoms, and some improvement in the paraplegia may generally be

anticipated, especially if an early diagnosis is made and acted upon. On the other hand, it is unusual to obtain a perfect recovery, particularly in regard to the vesical troubles. A patient may reach a stage in which he is able to perform all his ordinary duties, and even to walk many miles without great fatigue, but his control over micturition often remains very incomplete.

**Treatment.**—What has already been said with regard to the treatment of acute syphilitic myelitis applies to this more chronic type of lesion. The danger of bedsores and of cystitis is not nearly so urgent, and complete rest is rarely indicated. If the bladder trouble proves intractable, regular daily catheterization, with due precautions against sepsis should be instituted, and the patient taught to carry this out himself. A portable urinal is sometimes a convenience and occasionally a necessity.

### LANDRY'S PARALYSIS.

**History and Introduction.**—During the last half century this disease or its title, has enjoyed a chequered character. Every form of acute ascending paralysis which has borne the faintest resemblance to the case originally described by Landry has been called Landry's paralysis, whether the changes in the cord have been gross or insignificant, with the result that the literature has become hopelessly confused. E. W. Taylor says that "the tendency to consider cases of rapidly advancing paralysis, whether fatal or not, regardless of pathological findings, as Landry's paralysis, is very noticeable in much of the best recent work in America;" but this tendency is not confined to America, and may be found in any country of importance.

This unfortunate confusion can be traced to three principal sources. In the first place, many cases of acute poliomyelitis of the adult have been mistaken for Landry's paralysis during life, and the diagnosis has not been revised when examination of the spinal cord has revealed the inflammatory changes characteristic of the former disease. In the second place, the occurrence of acute cases of polyneuritis of unknown toxic origin has led observers to believe that Landry's original case, in which the peripheral nerves were not examined, was only an example of this condition. In the third place, acute ascending myelitis presents features which have provoked the diagnosis of Landry's paralysis in certain instances.

The writer desires to avoid any attempt, which can never be successful, to reconcile the various views which have been, and are, held upon this subject, or to classify the different morbid processes which have at one time or another carried the name of Landry's paralysis. It does not matter, and it will certainly never be known, what was the actual disease from which the cases recorded by Landry suffered. It is sufficient for our purpose that he called attention to a class of case in which the symptoms were those of a rapidly spreading motor paralysis, without atrophy or electrical changes in the muscles, with slight sensory phenomena and no sphincter disturbances, and in which he was unable by the methods at his disposal to detect anatomical changes. Does this class

of case still exist? The answer to this question is in the affirmative if allowance is made for improved histological technique, and if it is not forgotten that Landry would certainly have been able to detect the changes of acute poliomyelitis or any other acute inflammatory process in the spinal cord. It is clear, however, that "acute ascending paralysis" and "Landry's paralysis" cannot be regarded as synonymous terms; the latter is only one variety of the former.

It will be the object of this article to describe a morbid condition, which corresponds clinically to Landry's cases, in which the anatomical changes in the spinal cord would certainly have escaped notice with the methods of fifty years ago, and which presents no grounds for its inclusion in the group of the neuritides. There is nothing to be gained from imitating the course adopted by some writers, who describe a central and a peripheral form of Landry's paralysis.

**Etiology and Bacteriology.**—There are but few points of etiological importance, if we exclude those cases which should properly be classed under the term myelitis. A recent history of some infectious disorder, such as gonorrhœa, influenza, or typhoid fever, is sometimes forthcoming, but not with sufficient constancy to merit attention. In the majority of instances the disease attacks healthy adults, and the period of life between twenty and forty appears to be the most susceptible. Men are more often affected than women, but it is difficult to say whether the preponderance is great or insignificant. Exposure to extremes of heat or cold may be a predisposing factor. Seasonal or climatic influences are not known to play any part, nor is there any evidence of the occurrence of epidemics. During epidemics of acute poliomyelitis the earlier severe cases have often been mistaken for examples of Landry's paralysis until the opportunity for examining the tissues has revealed the true condition.

The bacteriology is in an immature condition, and it is not possible to say whether the disease is specific or whether it may occur as a result of various forms of intoxication, bacterial or otherwise. The large majority of cases in which spinal lesions have been absent or insignificant have given a negative response to bacteriological investigation. On the other hand, a few have provided findings of some interest. Roger and Josué recorded a case of acute ascending paralysis in which they found toxic changes in the cells of the lumbar enlargement. Sections stained for bacteria did not demonstrate the presence of any organism, but cultivation from the heart blood produced a diplococcus resembling the pneumococcus. Injections of the coccus into a mouse did not prove fatal, but a rabbit submitted to the same experiment died at the end of twenty-one days with symptoms of paraplegia, and the organism was recovered from its blood.

The writer investigated a case of the disease which was in the National Hospital under the care of Sir William Gowers with the following results: (1) A micrococcus was isolated in pure culture from the blood of the patient after death. (2) An organism indistinguishable from that which was cultivated was found in large numbers in the loose vascular tissue forming the external layer of the spinal theca. (3) A subdural injection

of the cultivated coccus into a rabbit produced after some days a rapidly spreading palsy. (4) The same organism was discovered in the theca of the rabbit, and isolated in pure culture from its blood. (5) In neither the patient nor the rabbit was the organism demonstrated in the spinal cord or the pia arachnoid, and in neither case were there inflammatory reactions in these tissues. Macnamara and Bernstein isolated from the blood and cerebrospinal fluid of a non-fatal case a coccus which bore some resemblance to that just described, but they were unable to obtain any positive results from experimental inoculations.

**Pathology.**—To the naked eye the central and peripheral nervous systems present little that is remarkable, with the exception of some general hyperemia of the cord. It is particularly to be noted that this organ is always firm and natural in consistence, provided that post-mortem decomposition has not taken place. On cross-section the vascularity of the gray matter may be noticed, and sometimes it is possible to detect small hemorrhages in its substance. The soft meninges present no evidence of exudation, serous or purulent.

The microscopic examination may, especially in cases which have run a rapidly fatal course, reveal practically no signs of disease beyond a few capillary hemorrhages, but if the Nissl and Marchi methods are employed a careful inspection will usually result in the discovery of the following changes:

1. **Cells.**—A smaller or larger number of the spinal cord cells, especially those of the anterior horns and of Clarke's column, present either early pericentral chromatolysis, or more or less complete loss of chromatin granules and excentration of nuclei. The most intense cell change is found in those parts which, judging from the clinical symptoms, were earliest affected; in the majority of cases the legs are the first limbs to fail, and the most marked cell changes are detected in the lumbosacral enlargement.

2. **Myelin.**—The myelin sheaths of the spinal cord nerve tracts, and to a less extent those of the peripheral nerves, often present a form of diffuse fatty change. In longitudinal sections the small droplets of fat are seen lying singly, or two or three together, along and between the nerve fibres, and do not fill the transverse area of the myelin sheaths. This appearance may be found in toxic states unassociated with paralysis, and does not therefore indicate necessarily any alteration of function on the part of the nerve fibres.

3. **Neuroglia and Vessels.**—There is no evidence of neuroglial proliferation, although some of the cells may appear to contain more protoplasm than normal. The vessels are engorged, but are free from changes in their walls or perivascular sheaths. Very rarely a slight excess of small round cells may be seen in the immediate neighborhood of one or two vessels.

The above are the only morbid changes found in most cases at the time of death. Occasionally, when the fatal termination has been postponed, the Marchi method will reveal the presence of true Wallerian degeneration in the spinal cord and peripheral nerves, probably secondary to the cell changes. Early fatty changes in skeletal muscles are often

detected, but definite atrophy is not present until some weeks after the onset.

Outside the neuromuscular tissues, the most constant findings have been an enlarged spleen, enlarged mesenteric glands, and some evidence of pulmonary or pleural complications, generally of a secondary character.

**Symptoms.**—There is frequently a distinct prodromal stage lasting some hours, days, or even weeks, during which the patient may complain of various subjective sensations. Pain in the back is not common, although it does occur, but pricking, tingling, numbness, pins and needles, are usually described and referred to the peripheral parts of the limbs. A general sense of languor or fatigue and a feeling of heaviness in the legs and arms may also precede any definite loss of power.

The development of *paralysis* takes place somewhat rapidly and smoothly or by definite stages. In the latter case there may appear to be stationary periods followed by rapid involvement of fresh areas. Usually the legs are affected first, but occasionally the arms and rarely the cranial nerves present the earliest signs. The paralysis spreads from one part to another, following roughly the lines of spinal innervation. The lower intercostals, for instance, are weakened before the diaphragm is involved if the disease is following an ascending course, and the peripheral parts of the arms are affected before the muscles of the shoulder girdle. When a whole limb has become paretic, however, the proximal muscles are just as powerless, or sometimes even more powerless, than those moving the hands or feet. The writer has on more than one occasion seen a patient lying perfectly helpless as regards any gross movement of the trunk or legs, yet able slowly to flex and extend his toes. In this respect the condition affords a most striking contrast to many cases of multiple neuritis, in which it is the general rule to find the peripheral muscles more paralyzed than the proximal.

From the beginning and throughout its course the paralysis is of the *flaccid* type, the muscles lacking their normal tone and presenting no opposition to passive movements of the limbs. No atrophy or diminution in electrical excitability can be detected in any muscle until two or three weeks after the onset, and even then the wasting is slight and the electrical alterations rarely exceed a lessened response to the faradic current.

If the motor paralysis continues to ascend, the movements of the head, and later those of the tongue, larynx, pharynx, and palate, may be interfered with, but it is unusual for a patient to survive sufficiently long after the respiratory musculature has been seriously embarrassed to develop marked cranial nerve symptoms. The loss of the accessory muscles of respiration, difficulty in swallowing and in articulation, are the principal phenomena brought about by the upward extension of the disease. The complete paralysis of all trunk as well as all limb muscles renders the patient particularly helpless.

In contrast to the severity of the motor symptoms everything else appears almost insignificant. The constitutional disturbance may be slight or even absent, and the temperature rarely exceeds 100° or 101°, unless it rises rapidly within a few hours of death. The intellect and memory remain clear so long as respiration is effectually carried on,



and, in spite of dyspnoea, the patient is usually cheerful and sanguine concerning his condition. The paresthesia associated with or preceding the onset may continue for some time, and, in addition, complaint is often made of cramp-like pains in the immobile limbs, which may be relieved by change of posture.

Tests for detecting loss of sensibility in the skin or deep tissues usually discover no change from the normal, unless it be a slight dulling in the cutaneous areas below the knees. On the other hand, a suggestion of muscle tenderness may be elicited by firm pressure. The sense of passive movement and of position remains unaffected. Every deep or superficial reflex disappears in the paralyzed parts, and is usually unobtainable as soon as any decided paresis has developed. The plantar response remains flexor in type so long as it can be elicited at all.

The rectal and vesical sphincters are unaffected, although, in consequence of the paralysis of the abdominal muscles, some difficulty may be experienced in emptying both the rectum and bladder. Incontinence does not occur, except as the result of a distended bladder, and the patient is aware of the natural calls to defecation and micturition. With ordinary care decubitus can be avoided, as there is no special tendency to the formation of trophic sores nor any deficiency in healing power. Vasomotor disturbances are slight, if present at all; the skin over the paralyzed parts is moderately moist, at any rate in the early days. A *tâche* may sometimes be elicited.

The pulse may continue of fair strength and regular rhythm long after respiration has become difficult. With a fatal termination respiration gives out for a considerable time before the circulation fails, and artificial respiration may often preserve life for many hours. If life is prolonged the most serious complications are pulmonary and bronchial catarrhs, with which the patient is hardly fit to cope.

When neither the disease itself nor any complication proves fatal, the patient then enters upon the stage of recovery, and this presents some features of interest. The muscles often show a moderate degree of general wasting within three or four weeks of the onset, but the atrophy is diffuse, and not limited to particular groups. The response to faradism may be diminished, but is very rarely lost, and the galvanic current produces a normal or slightly sluggish contraction.

Recovery in power and in nutrition takes place slowly and equally over the whole of the musculature, and it is rare for any particular set of muscles to hang behind the others. Occasionally those of the limbs do not respond to treatment as quickly as those of the trunk. Contraction and deformities are rarely seen, except as the result of unskilled attention. Remissions and recurrences are not known to occur.

**Prognosis.**—Owing to the confusion which has existed, it is impossible to give statistics throwing any valuable light upon the mortality. In the opinion of the writer Landry's paralysis is a very fatal disease, and in this respect contrasts unfavorably with acute toxic polyneuritis and also with acute poliomyelitis.

At the same time it is undoubtedly true that there are instances of true Landry's paralysis which do end favorably, and in such there is

no reason to expect anything but an exceedingly good or even perfect recovery of function. As will be readily understood, the gravity of any particular case depends almost entirely on the condition of the respiratory musculature. So long as the diaphragm and the accessory respiratory muscles are intact, or at any rate capable of considerable work, there is, in the absence of complications, no serious cause for anxiety. When the diaphragm and intercostals are both very feeble, and the respiration is being carried on largely by the sternomastoids and the other neck muscles, the appearance of cyanosis, the look of distress, and the fogging of the patient's intellect are danger signals of the gravest import. The most favorable cases are those in which, although muscular weakness is very general, the loss of power is nowhere complete. Pulmonary and bronchial troubles must always be regarded as serious.

**Diagnosis.**—This is of importance not so much from the point of view of treatment (although it may not be many years before a correct diagnosis may be essential for this also) as for the purpose of giving an accurate prognosis while the disease is still in its acute stage.

When a patient is suddenly seized with an acute form of paralysis involving a large part of his musculature, two questions may well be asked by his friends: Is he going to live? If he lives, will he be permanently disabled? The medical attendant will ask himself, "Is this a case of acute ascending myelitis, of acute poliomyelitis, of acute toxic polyneuritis, or of Landry's paralysis?" Upon the answer to this question must the prognosis largely depend. For instance, it may be stated without hesitation that the diagnosis of acute toxic polyneuritis renders the prognosis bright as regards both life and fair recovery of power, that of acute poliomyelitis, on the other hand, hopeful as regards life, but very grave as regards return of activity.

There should be no difficulty in distinguishing acute ascending myelitis from Landry's paralysis on account of the severe sensory loss and sphincter disturbance of the former condition, although the spread of motor palsy is often similar in the two diseases. The mistake of calling a case of acute and widespread poliomyelitis one of Landry's paralysis has been often made, but attention to the following general principles will assist toward a proper diagnosis.

1. *The constitutional symptoms* in the severe cases of acute poliomyelitis are more marked than those in Landry's paralysis, the range of temperature is higher, often reaching 102° to 104°, the malaise and anorexia more profound, vomiting and disturbance of the alimentary canal more frequent. In children convulsions are often associated with the former ailment. The older the patient the less likely is acute poliomyelitis to be present. Cases of acute toxic polyneuritis may, in many instances, run their course with no corresponding features.

2. *Sensory Phenomena.*—In Landry's paralysis there may be, and often is, complaint of pain in the back, of sensations of numbness and tingling in the extremities before and during the onset of paralysis. The only sensation complained of in the paralyzed limbs is that of discomfort or of cramps, arising, partly at any rate, from their immobility; they may be handled without giving rise to anything more than slight tenderness

on deep muscular pressure. In acute poliomyelitis the pains complained of are more urgent, and affect not only the back, head, and neck, but frequently the limbs as well. Paresthesia may be present, but the more prominent feature is the pain, sometimes very severe, elicited by passive movements of the affected extremities. In both diseases cutaneous sensibility is usually unimpaired. In acute toxic neuritis, numbness, pins and needles, and sharp pains are common in the extremities, and very marked tenderness of muscles, sometimes of nerves, is the rule. There may or may not be relative anesthesia in the glove and stocking areas.

3. *Motor Phenomena.*—In both Landry's paralysis and acute poliomyelitis the paralysis is flaccid and may be general. The escape of a single muscle or of a group of muscles in a region where all the others are affected, or a marked asymmetry in the condition of corresponding muscles on the two sides of the body, is suggestive of acute poliomyelitis rather than of Landry's paralysis. Within a few days or a week of the onset in cases of acute poliomyelitis, if death has not occurred, it is usual to find rapid recovery in some parts and early atrophy and electrical changes in others. In Landry's paralysis recovery, if it takes place, is slow and evenly distributed, without marked atrophy.

In acute toxic polyneuritis the limbs are more affected than the trunk, and the peripheral parts of the limbs more than the proximal. Atrophy and electrical changes, perhaps only slight, quickly make their appearance, especially in the dorsiflexors of the ankles and extensors of the wrists. In the same disease, the most marked incidence is generally upon the diaphragm, in which case respiration becomes entirely costal. Of the cranial nerves, those supplying the facial muscles are often picked out and may be the only ones to suffer. Occasionally the palatal or ocular muscles are affected. In Landry's paralysis the musculature is affected generally and evenly, the trunk and limbs presenting a degree of paresis which is more or less symmetrical and equal. In cases in which the paralysis first affects the lower extremities it is not uncommon to find these parts most paretic at the time of observation, but the spread of the disease will then be a progressive one in an upward direction, and the thoracic muscles may succumb before the diaphragm, or both may be equally weak. When the muscles innervated by the cranial nerves are involved, deglutition, phonation, articulation, and more rarely the movements of the face and jaws, may be impaired, but the fatal termination generally takes place before any of these actions are abolished. The condition may be progressive over two, three, or more weeks, and yet be unassociated with any definite local atrophy of muscles.

4. The *deep reflexes* are abolished in the affected parts in Landry's paralysis and toxic polyneuritis. The abdominal reflexes are often retained in acute toxic polyneuritis and nearly always absent in Landry's paralysis. The plantar reflexes are absent or flexor in type in both instances, and the sphincters are only temporarily, if at all, disturbed.

In addition, it is interesting to note that relapses and recurrences are not infrequent in toxic polyneuritis, although they are extremely rare or unknown in poliomyelitis and Landry's paralysis.

**Treatment.**—No measures are known to have any definite influence on the course of Landry's paralysis. The patient must be placed at complete rest, the head slightly raised, and changes of posture allowed for the sake of comfort. There is little liability to the formation of sores provided ordinary care and cleanliness are exercised. The bladder may require catheterization in the first day or two. A purge should be given, and if there is any fever a diaphoretic mixture is indicated. Every effort must be made to prevent the excitement of bronchial catarrh by exposure to cold, and the administration of atropine or belladonna, together with strychnine, may relieve the respiratory distress by diminishing bronchial secretion when the intercostal muscles and diaphragm become involved. Oxygen and artificial respiration may prolong life, although the necessity for their use is generally a token that a fatal termination is impending.

No active treatment of the paralyzed parts is indicated until the dangerous stage has passed and the disease has definitely taken a favorable course. The employment of massage and electricity may then tend to hasten the recovery of power. Passive movements should be carried out daily in order to prevent arthritic adhesions.

### ABSCESS OF THE SPINAL CORD.

Intramedullary collections of pus are very uncommon in the spinal cord as compared to the brain, and are not of great clinical importance. It is doubtful whether the fluid found in some of the early recorded cases was really pus or the result of necrosis following spinal thrombosis.

**Etiology.**—Spinal abscess occurs most frequently as a secondary complication of distant septic processes, especially those involving the urogenital system, the lungs, and the heart. Two cases observed by Cassirer arose in connection with a suppurating dermatitis and an infective endocarditis respectively.

In some cases, especially those of pulmonary origin, suppurative meningitis has been found in association with the central abscess, but it must not be too hastily assumed that the abscess was secondary to the meningitis. The frequency of suppurative meningitis and the rarity of spinal abscess are strongly opposed to this view. It is possible that the meningitis is sometimes a consequence of the abscess, and it is probable that in other instances the two conditions arise independently from a common cause. On the other hand, small abscesses are very occasionally seen within the spinal cord in cases of suppurative meningitis under circumstances which point to the dependence of the former upon the latter. In another class may be grouped instances of spinal abscess which have been grafted upon a cord already damaged by fracture, dislocation or caries or carcinoma of the vertebral column. Most of these must be regarded as secondary to a skin infection or to a purulent cystitis, the result of the initial lesion, the incidence of the metastatic process being determined by the evascularized and non-resistant condition of the spinal tissues at the seat of compression. Finally, a small focal

abscess is sometimes found in the midst of a non-purulent infective myelitis.

**Pathology.**—An abscess may be suspected when the cord presents a fluctuating swelling, especially if a purulent meningitis is apparent on the surface, but the organ must be opened in order to exclude a syringomyelic or hematomyelic cavity. The purulent collection is always more or less central, occupying a position at the base of the posterior horns or in the posterior commissure so much favored by extravasations of blood and by gliomatous cavities. Occasionally the white matter, usually that of the posterior columns, is encroached upon. The pus is of a greenish-white color, generally thick, and contains the remains of nervous tissue in the form of drops of myelin and fat. The abscess cavity may be focal or may extend through many segments of the cord. The walls in acute cases are friable; in more chronic cases a more definite lining may be formed by proliferated neuroglial tissue. Degenerative changes are found in relation to the nerve elements involved in the process.

**Symptoms.**—Little is known of the symptomatology for several reasons. The evidence of abscess formation is often obscured by the signs of meningitis or myelitis, or the suppuration is frequently a terminal event, and occurs at a time when the patient's general condition does not allow of careful investigation. The process may run an afebrile course, or may be associated with rigors and an irregular pyrexia. The spinal symptoms are those of a focal or diffuse myelitis, and will depend upon the site of the lesion. Flaccid motor paralysis, disturbance of sensibility and of the sphincters, loss or exaggeration of the tendon reflexes, form part of the clinical picture, but the patient rarely survives long enough for muscular atrophy to take place. Occasionally symptoms pointing to multiple purulent foci present themselves.

**Diagnosis.**—This is most likely to be arrived at in cases of pulmonary sepsis, although the symptoms may be ascribed to myelitis or meningitis, and the discovery of an intramedullary abscess on the postmortem table may come as a surprise. Metastatic abscesses in other organs are the chief indications of the spinal condition. In cases of compression paraplegia followed by cystitis, the upward spread of spinal symptoms may suggest the spread of a purulent focus, especially if it be accompanied by severe constitutional disturbance.

**Prognosis.**—This is invariably bad and a fatal termination not often long delayed.

**Treatment.**—This is only prophylactic and must be directed against the primary cause.

## VASCULAR DISORDERS OF THE SPINAL CORD.

**Anemia.**—There is no proof that a *general* anemia is capable of producing organic lesions of the human spinal cord, nor does our knowledge permit us to attribute any spinal symptoms to that source. On the other hand, experimental results justify the supposition that the most specialized nervous elements react quickly to alterations in the blood state, and conduce to the belief that disorders of nervous function may

result from anemia of the spinal cord, without the presence of signs of organic change.

In certain varieties of general anemia, particularly in pernicious anemia, it is not uncommon to find changes in the spinal cord, but it is highly probable that these are not the direct result of the altered blood state, and that they should properly be referred to the effect of some toxic agent, to which the anemia is also secondary.

The paraplegia which results from excessive loss of blood has no known anatomical basis, and it is difficult, therefore, to say which part of the nervous system is chiefly at fault. Speaking generally, it may be quoted as an example of the vulnerability of the nervous tissues to anemia, and the question of its cortical, spinal, or peripheral origin left for further investigation.

When the subject of *local* anemia is brought under consideration, there are more facts to guide us in forming conclusions. This may be partial or complete, and may have its origin in (1) thrombosis due to a blood state or to disease of vessels, (2) embolism, (3) strangulation of vessels by pressure of tumors, thickened meninges, or displaced bone, and (4) vasomotor constriction or spasm, or (5) dilatation.

1. Little is known about *thrombosis* as the result of an abnormal blood state in the spinal cord, unless some of the changes in toxemic conditions can be accounted for in this way. Arteriosclerosis of the spinal vessels is by no means uncommon, but for some reason which is not obvious spinal thrombosis is far less common than cerebral thrombosis. Probably some of the cases which are usually termed senile paraplegia or senile myelitis have a pathological basis founded on softening of arteriosclerotic origin. If this is so, their symptomatology is more or less identical with that of cases of syphilitic myelitis. Arteritis due to syphilis, and more rarely to tubercle, is the most potent and frequent cause of thrombosis.

2. *Embolism* in the spinal cord is a rare event, but cases have been described by von Leyden, Weiss, and Gowers which bear this interpretation. In all these instances the patients were suffering from mitral disease, and in two of them the postmortem examination showed the presence of spinal softenings. Experimental spinal embolism has been successfully produced by means of foreign particles introduced into the lumbar arteries. Hoche showed that foci of simple softening were generally found in the central gray matter, owing to the larger caliber of the anterior spinal artery. On the other hand, the use of irritating vegetable seeds resulted not only in simple necrosis, but in an inflammatory reaction. The animals operated on became rapidly paraplegic.

3. Spinal vessels are frequently narrowed or occluded by the pressure of tumors, thickened meninges, or displaced bone, with the result that areas of tissue are evascularized and sooner or later become necrotic. The term "compression myelitis" was formerly used to denote the condition.

4. Vasomotor constriction or spasm of the spinal arteries is one of those phenomena the occurrence of which may be regarded as possible, but the proof of which is not yet forthcoming. Paroxysmal paraplegia or

monoplegia in a person who presents no symptoms of spinal disease between the paroxysms may be interpreted on the hypothesis of vasomotor spasm, and a similar construction placed upon cases of intermittent paresthesia of the extremities, but these are matters of speculation rather than of demonstrated fact. Vasodilatation can certainly be instrumental in slowing the blood stream, and so producing a relative anemia and possibly a thrombosis. Such an event may be the result of the vasomotor paralysis produced by toxic agents, and a special form of paralytic vasodilatation has been described with syphilitic myelitis.

The *effects of local anemia* are dependent upon the degree to which the cutting off of the blood supply attains. A diminution in the blood stream may exert deteriorating influences upon the nerve cells of the anterior horns, while the rest of the tissues escape. A further diminution may destroy the myelinated nerve fibres, including their axis cylinders, the neuroglia still maintaining sufficient integrity to undergo proliferation, and thus to inaugurate a process of repair and sclerosis. Finally, when the evascularization is complete all tissues succumb in the affected area, and, according to Schmaus, the attempts at repair and the removal of debris are carried out by mesoblastic elements, with the result that a connective tissue scar, with or without the formation of a cyst, replaces the parts which have been destroyed. In this respect the effects of focal thrombosis in the spinal cord resemble those which obtain in the brain, although the common etiological factors are not the same.

**Hyperemia.**—Hyperemia of the spinal cord at one time enjoyed a position of considerable importance in relation to the pathology of spinal diseases. Nowadays it has fallen from its pedestal, and, whether active or passive, is regarded rather as a concomitant phenomenon of physiological or morbid activities than as a primary factor capable of evoking clinical symptoms or anatomical lesions. It is sufficient to say that the spinal cord shares with other organs the power of attracting an excess of blood during functional activity and the inability to avoid plethora when it is the seat of inflammation or of passive congestion.

### HEMORRHAGE INTO THE SPINAL MEMBRANES.

**Extradural Hemorrhage.**—Extravasation of blood may occur between the dura mater and the periosteum lining the inner surface of the vertebræ. This space contains, especially posteriorly, a considerable quantity of loose areolar and fatty tissue, in which lies a rich plexus of veins. Bleeding into this epidural region occurs in the large majority of cases as the result of trauma, fractures of the vertebræ, with or without laceration of the dural sheath, being the most common cause. Less frequently extravasation may be due to violent involuntary muscular contractions, such as those which are associated with tetanus, eclampsia, and infantile convulsions. The venous congestion of cardiac and pulmonary disease combined with the excessive action of the respiratory musculature in terminal dyspnoea may also be responsible.

Finally, the bursting of an aortic aneurism into the extradural space will in rare instances produce a hemorrhage of a gross character. With

the exception of the latter cases, the amount of blood poured into the extradural space is rarely sufficient to produce symptoms, and its almost invariable occurrence within a few hours of death from other causes renders these symptoms likely to be overlooked. Only in exceptional instances, such as with aneurism, is pressure upon the spinal cord likely to be brought about. These hemorrhages are therefore usually recognized only postmortem. The blood may be limited to a small area or diffused throughout the length of the vertebral column, sometimes penetrating, with the spinal nerves, the intervertebral foramina.

**Intradural Hemorrhage; Hematorrachis.**—In this the blood is effused into the arachnoid sac and is more or less mingled with the cerebrospinal fluid, to which it gives a hue proportionate to the size of the extravasation. As a general rule, the source of bleeding is in the vessels with which the pia mater is richly supplied. In other cases blood has made its way into the spinal theca from the cranial cavity, where it has escaped as the result of a hemorrhage of traumatic or non-traumatic origin. Laceration of the dura mater may also allow of extravasation into the arachnoid sac from surrounding tissues. Aneurism of a vertebral artery is a very rare cause of intradural hemorrhages. The most common origin of small hemorrhages or ecchymoses on the surface of the pia is to be found in the fulminating forms of septic meningitis. In such cases numerous extravasations, sufficient to color the cerebrospinal fluid, are not infrequent. Hemorrhages are also met with in connection with tetanus, eclampsia, and epilepsy, as well as with purpura, scurvy, and hemophilia. Urgent dyspnœa as a result of pulmonary or cardiac disease or of respiratory paralysis is an occasional causal factor.

In most cases there are no symptoms referable to the effusions of blood which can be distinguished from those of the primary lesion, whether it is of cerebral or spinal origin. In other instances, usually of traumatic, but very occasionally of spontaneous origin, to which the term spinal apoplexy has been given, the amount of blood effused is sufficient to produce a recognizable symptom complex.

**Symptoms.**—These are produced by pressure upon the leptomeninges and upon the spinal roots, and are characterized by the rapidity of their development. Sudden, violent pain referred to the back, associated with tenderness over a considerable length of the vertebral column, is the first indication. This may give rise to spinal rigidity or opisthotonos, and is quickly followed by root pains in the distribution of the spinal nerves, by various paresthesias in the extremities, and by involuntary spasmodic contractions in the muscles of the trunk, legs, and arms. Pain may be evoked by attempts at micturition and defecation or there may be temporary retention of urine and feces.

Such are the symptoms of meningeal and root irritation. They may be short-lived or they may merge into more serious disturbances. Corresponding to the site of the hemorrhage, paralysis of the arms or legs may develop, and hyperesthesia of certain cutaneous areas may be followed by anesthesia and analgesia, both superficial and deep. This is particularly liable to affect the lumbar and sacral root areas, owing, probably, to the collection of blood in the lowest part of the thecal sac and conse-



quent pressure upon the cauda equina. Abolition of the tendon reflexes and retention or even incontinence of urine and feces may be observed.

The evolution of this clinical picture is usually associated with a certain degree of shock and in severe cases with loss of consciousness. The pupils are contracted at first and may be dilated later. The pulse is small and rapid, and there may be an initial fall of temperature.

The disease reaches its height in the course of a few hours or a day or two, and it is during this time that life is most dangerously threatened either from shock, respiratory paralysis, or in very rare instances from extension of the hemorrhage into the cranial cavity. An early retrogression of symptoms may be expected in those who survive, but the period of blood resorption is often attended by an exacerbation of the phenomena and by pyrexia and constitutional disturbances. This is not often fatal, although some days may elapse before definite evidence of progressive recovery from paralysis is forthcoming. From this time onward complications arising from decubitus or cystitis are the chief source of anxiety, and these may generally be successfully dealt with provided the spinal cord itself has not sustained irreparable damage. A considerable degree of recovery is generally attained in the course of six or eight weeks, but residual troubles in the form of atrophic palsies, bladder disorders, and paresthesias may require many months to clear up, or may even be permanent in the worst cases.

**Prognosis.**—This has already been indicated by reference to the chief sources of danger. The nature of the primary lesion in traumatic cases, and particularly the level of the cord, to which the symptoms point, must be taken into account. The confinement of symptoms to those of meningeal irritation is a favorable sign and generally denotes a rapid recovery. Signs of an intramedullary lesion render the outlook very grave from the point of view of recovery from paralysis and sphincter disorders.

**Diagnosis.**—The diagnosis is rendered easy, as a rule, by the history of trauma immediately preceding the development of symptoms of meningeal irritation. A slow evolution of the latter without pain is a very exceptional occurrence, although it has been recorded. When there is no history of injury, meningitis may be suspected, and the differentiation will then depend on the slow onset of the latter condition, the more pronounced early pyrexia, and especially on the examination of the cerebrospinal fluid which may be blood-stained in either case.

The diagnosis of hematorrachis from hematomyelia depends chiefly upon the early preponderance of paralytic over irritative phenomena and the more localized character of the pain in the latter disease. It must be remembered that intradural and intramedullary hemorrhage may co-exist in traumatic cases.

Gowers refers to the possibility of mistaking spinal apoplexy for strychnine poisoning, and notes the case reported by Dixon, in which an extensive hemorrhage into the dural sac gave rise to violent paroxysms of muscular spasm and general pain. In such instances the question as to whether an effusion of blood into the arachnoid space is the result or the cause of muscular spasms may often be difficult to answer.

**Treatment.**—The first essential treatment is physical and mental rest, which is best promoted by placing the patient in bed and administering a hypodermic injection of morphine. It is generally assumed that the prone position exerts a favorable influence on hemorrhage or inflammation in the spinal tissues, but on what grounds this opinion is based it is difficult to see. The importance of securing the patient's repose and comfort probably outweighs any advantage that can be gained from invoking the help of the laws of gravity. The application of ice to the spinal column may be of service in arresting hemorrhage. Scarification of the surface over the spine at the seat of pain is suggested by Gowers as probably the wisest treatment at the onset. Calcium salts may be given by the mouth in order to avert further oozing, although they can hardly exert any influence on the initial hemorrhage.

The after-treatment is that of atrophic palsies in general, and the persistence of symptoms indicating pressure on the spinal cord or cauda equina may eventually justify an exploratory laminectomy.

### HEMATOMYELIA (HEMORRHAGE INTO THE SPINAL CORD).

Hemorrhages within the substance of the spinal cord are not infrequently found postmortem in cases of spinal inflammation or spinal thrombosis, but the symptoms of these cases belong to the primary disease, and are not those of a true hematomyelia. Capillary hemorrhages are also common in patients who have died with urgent dyspnoea, particularly in cases of pulmonary disease, myasthenia gravis, and in all instances of paralysis of the respiratory musculature. Here, again, there are no clinical symptoms referable to the hemorrhages. The term hematomyelia is reserved for cases in which a hemorrhage is mainly responsible for the clinical picture.

**Etiology.**—Since sclerotic vascular change is one of the most important factors in the production of cerebral hemorrhage, it might be expected that similar changes in the spinal cord would determine the occurrence of spinal hemorrhage, but this is not the case. Sclerotic degeneration of spinal arteries is not uncommon, but it rarely gives rise to hematomyelia, and the comparative rarity of the latter condition as compared to cerebral hemorrhage is sufficient evidence of the essential difference in their etiology. Possibly the circuitous course of the spinal vascular supply modifies the effects of high arterial tension.

By far the most common factor is trauma or strain, although there are a few cases in which hemorrhage appears to have occurred without any obvious cause or predisposition. Included under the term trauma are cases of injury to the spinal column, with or without fracture or dislocation, cases of concussion due to falls on the head, on the feet, or upon the sacral region. Obstetrical injuries have also caused hemorrhage into the child's spinal cord. The strain of childbirth and that of great muscular exertion may give rise to the condition, and excessive coitus has been cited by Gowers as a possible causal factor. Hemophilia, congenital or acquired fragility of vessels, purpura, and other rare conditions have been assumed to exert an influence.

Hematomyelia may occur at all *ages*, but is most common between twenty and forty years, the period of greatest physical exertion and exposure to injury. The laborious nature of their occupations renders men much more liable than women to the disease. The incidence of hemorrhage upon the gray matter is generally explained by the rich vascular supply of that tissue and the comparative looseness of its texture.

**Pathology.**—In recent cases the appearance of the cord may be normal or the soft meninges may present some ecchymoses. Palpation will often detect a soft fluctuating swelling, and, if the hemorrhage is extensive in its transverse direction, the dark bluish-red color of the blood clot may be visible through the surrounding white matter. In more severe cases a large part or the whole length of the cord may form a tubular thin-walled, blood-containing sac. The most common site is the cervicodorsal enlargement; its occurrence in the dorsal or lumbosacral region is rare.

A series of transverse sections shows that the extravasation may be limited to a segment or two, when it will have a round or oval shape, or it may extend through many segments in the form of tapering prolongations upward or downward, in which case it will have from the vertical aspect a more or less spindle-shaped contour. Closer scrutiny reveals the limitation of the blood to the gray matter at most levels, although at the seat of the original leakage the white matter may also be seriously encroached upon and torn up. The track pursued by the hemorrhage usually involves the bases of the posterior horns, but extends also into the anterior and lateral gray substance.

Multiple foci are relatively often seen (in 10 out of 32 cases according to Doerr). The color depends on the age of the hemorrhage, red in the early cases, it becomes brown or ochre in those of longer standing. In very old cases the site may be marked by a serous cyst, or more commonly by longitudinal cracks or fissures with well-defined walls in the posterior gray matter. It is stated that the blood rarely courses along the central canal.

**Histology.**—The nerve substance surrounding the blood in recent cases is seen to be partially disintegrated by mechanical means and by the secondary oedema. Proliferation of the neighboring neuroglia soon takes place, and granular cells are seen in considerable numbers. These appearances have been regarded as inflammatory, but this is not correct if the word is used in its ordinary sense, and it seems a pity to assume a secondary myelitis in order to describe the process of repair. Secondary changes in the nervous elements comprise disappearance or atrophy of ganglion cells, degenerations in the ascending and descending spinal tracts, and atrophy of the anterior root fibres.

**Symptoms.—Onset.**—Only in rare cases is this preceded by warning in the form of tingling or numbness corresponding to the site of the future lesion. As a rule, the development of symptoms is absolutely sudden, exceptionally occupying a few minutes to half an hour. A slower onset is suggestive of a secondary hemorrhage, except in those cases when a slight leakage is followed shortly by a more extensive extravasation.

Abrupt *paralysis* involving all parts below a certain level is character

istic of the disease, and may or may not be accompanied by pain. The latter, when it is present, is often of an intense radiating character, or may be described as a burning sensation constricting a part of the trunk, usually the upper thoracic region. Consciousness is usually retained, except in the most severe hemorrhages, and no disturbance of general health is immediately noticeable. If the lesion occupies the cervical *enlargement*, examination at this period reveals a flaccid palsy of all four extremities, together with paralysis of the thoracic and abdominal muscles. Unless the fourth cervical segment has been involved, the diaphragm will continue to act. In addition to the motor paralysis complete retention of urine and feces supervenes. Interruption of the oculopupillary fibres leaving the cord at the level of the eighth cervical or first dorsal segments produces contraction of the pupils and narrowing of the palpebral fissures. Splanchnic palsy entails distension of the abdomen, which may add to the respiratory embarrassment occasioned by the absence of intercostal action. The pulse may be slowed or unaltered. The temperature usually remains normal at first, and rises after a day or two.

In the course of a few days, unless a fatal termination has ensued, a marked retrogression of symptoms usually takes place. The arms remain flaccid and present evidence of muscular atrophy, associated with changes in the electrical reactions. The lower extremities, on the other hand, become spastic, with increased tendon reflexes and extensor plantar responses. A certain amount of recovery in power is often exhibited at this stage both in the trunk and legs, but the abdominal reflexes remain absent. Incontinence of urine sets in and obstinate constipation, alternating with incontinence of feces, may be expected. Priapism has been noted, but is not always present.

Cutaneous sensibility may remain generally impaired, but more frequently tactile stimuli become recognizable, while painful and thermal sensibility are still lost. Unless great care has been exercised bedsores may have rapidly developed in the first few days.

Occasionally the symptoms quickly assume, or have from the beginning, a unilateral character corresponding to what is known as Brown-Séquard's paralysis. In that case one arm may present flaccid, and the corresponding leg spastic, paralysis, painful and thermal sensibility being lost in the affected arm and in the trunk and leg of the opposite side.

Hemorrhage into the *dorsal cord* produces a similar picture, with the exception that the arms are unaffected and the oculopupillary fibres escape. When the *lumbosacral enlargement* is the site of disease the lower extremities are rendered powerless, all their deep and superficial reflexes are abolished, and the sphincters are relaxed and incontinent. In these rare cases the retrogression of symptoms is usually less marked, although a few muscles may regain power and complete sensory loss may be replaced by that of the dissociative type in certain areas.

With hemorrhage into the *conus medullaris* there is paralysis of the bladder and rectum, impotence, and anesthesia of the perineal region, the anus and the genital organs, occasionally of the posterior surface of the thighs. Vasomotor and sensory phenomena may be observed in

paralyzed regions, early vasodilatation and hyperidrosis being succeeded by a pale dry skin in the later stages.

The future course of the severe cases described is generally marked by continued improvement up to a certain point, but some muscular atrophy usually remains when the cervical or lumbar enlargements have been the site of hemorrhage, and some degree of spastic paraplegia when the cervical or dorsal regions have been the parts affected. In less extensive hemorrhages the injury may be confined to one side of the gray matter, when sensory and motor phenomena are limited to a single limb. More rarely an area of motor paralysis is observed without any sensory loss, or even a partial hemianesthesia without any degree of paresis. These mild cases may recover completely, although the occurrence of some muscular atrophy usually precludes such a favorable outlook.

**Diagnosis.**—Hematomyelia is, as a rule, easily distinguished from other forms of spinal disease by reason of its abrupt onset and the rapid partial amelioration which supervenes on the resorption of extravasated blood.

*Hematorrachis* or meningeal hemorrhage is characterized by more diffuse pain along the whole length of the spine, by the preponderance of signs of irritation over those of paralysis, and by the still more rapid retrogression of symptoms. Lumbar puncture may aid in the diagnosis, but it is contra-indicated in cases of hematomyelia. *Acute infective myelitis* is associated with marked constitutional disturbance and pyrexia, and a less rapid onset of symptoms. *Syphilitic myelitis* may have a very abrupt development, but the history of lues and the premonitory symptoms are usually sufficient for a correct diagnosis.

In children hematomyelia may simulate an *acute poliomyelitis*, and in certain cases the differentiation may conceivably be impossible. The almost invariable occurrence of sensory and sphincter disturbances in the former and the pyrexia and malaise associated with the latter disease are safe guides in the majority of instances.

The clinical picture of a case of hematomyelia may be identical with that of *syringomyelia*, in which circumstances the diagnosis can only be arrived at from a consideration of the history of onset and the absence or presence of evidence of progressive symptoms.

**Prognosis.**—Life is threatened at the onset when the extravasation is at a high level or when it is severe and continuous. A few cases die rapidly, either from respiratory failure or from shock. The majority survive the early dangers, and the future outlook then depends upon the avoidance of pulmonary complications and of sepsis in connection with bedsores or cystitis. Complete recovery from paralysis can hardly ever be expected, and the prospects in this connection must be gauged from the atrophy and the electrical reactions of affected muscles. So long as a muscle responds to the faradic current the hope of its regeneration may be maintained. The disappearance of all response to electrical stimuli is a sure sign that the paralysis is permanent. The parts affected by spastic paresis may reasonably be expected to show continued improvement over a long period of time because the encroachment of the disease upon the white columns is usually only temporary and incomplete. The return of sphincter control depends upon much the same

anatomical data, except in cases of lumbosacral hemorrhage, in which the sphincters are more likely to be permanently paralyzed.

**Treatment.**—It is imperative to secure absolute rest. The patient should be placed in bed, on a water mattress if possible, and given a dose of morphine hypodermically. The prone position is generally advised, but is probably quite unnecessary and certainly not conducive to the patient's repose or comfort. Ice may be applied to the back. The bladder and skin should from the very beginning receive assiduous care and attention. The bowels must be relieved by aperients or enemata, or both. The patient should be warned that coughing, sneezing, and all movements are injurious, and his diet should be nutritious and non-stimulating. Insomnia may be treated by bromides, veronal, sulphonal, or paraldehyde, but the administration of sedatives must be regulated by the condition of the respiratory muscles and organs.

Active treatment of the paralyzed parts should not be undertaken for six or eight weeks after the last sign of hemorrhage has been observed, and the first form allowable is electricity, which entails no active movements. This may be followed by massage and passive movements, and still later by gymnastics, baths, and other remedies devised for those who have faith in them. Iron, quinine, and arsenic are preferable to strychnine and digitalis in the way of tonics, and a change of air and scene is probably of most value to a patient who has had a prolonged period of dull inactivity and confinement.

The treatment of the cystitis and bedsores, which are sometimes inevitable in spite of all precautions, is the same as of those of other origin, if it is remembered that every care must be taken not to move the patient more than is absolutely necessary in the early days of the disease.

## CHAPTER VII.

### TOPICAL DIAGNOSIS OF DISEASES OF THE BRAIN.

By JOSEPH COLLINS, M.D.

**Introduction.**—There are certain areas of the cerebral cortex which have definite, highly specialized function, to which the name *centres* has been given. Thus, we speak of the motor centre, the centres for the different varieties of sensation, and of the speech centres. Some of these areas are more closely delimited than others, such, for instance, as the motor area. The determination of the areas of the cortex, to which highly specialized function is confined, was one of the brilliant contributions to medicine of the nineteenth century.

Topical diagnosis of brain disease is possible only when focal symptoms exist. The so-called general symptoms of disorder of the brain are of great importance in leading to a diagnosis of disease of the brain, but they are of no value in determining the part that is diseased. The *focal* symptoms are, first, those that manifest themselves through the emissive channels, twitching, spasm, convulsion, paralysis, dysarthria, anarthria, and second, those that manifest themselves through the percipient, apperceptive, and interpretative mechanism, such as disorder of tactile, thermal, pain, postural and spatial sensibility, apraxia, agnosia, astereognosis, aphasia, agenesis, and anopsia.

The localization of diseases within the substance of the brain is dependent upon the existence of symptoms similar in a measure to those resulting from disease of the cortex, because they are due to lesions of pathways leading to and from the cortex. In addition, there occur symptoms due to involvement of parts having more or less specific function, such for instance as the nuclei of some of the cranial nerves.

**The Motor Area of the Brain-cortex.**—The centres for voluntary movement are situated in the anterior central, precentral, or ascending frontal gyrus. This gyrus extends on the mesial surface of the brain from the paracentral lobule to the operculum. It was to this area of the cortex that Fritsch and Hitzig first attributed representation for voluntary movements, and though a vast amount of clinical and experimental evidence was offered to show that motor representation was not narrowly confined to this area, yet the preponderance of the evidence today indicates that motor representation is confined essentially to this region.

Many workers have conclusively shown that true motor responses are never obtained save from stimulation anterior to the Rolandic fissure. To different parts of this anterior central convolution are allotted centres for the different parts of the body. It may be said that a manikin, with its head resting on the operculum and its feet over the paracentral lobule, will adequately convey the representation, *i. e.*, the centre for the face

and its components occupies the lower third, that for the arm and the trunk the middle third, and that for the legs and feet the upper third. Formerly it was thought that there was considerable localization of function to the mesal surface of this area, the paracentral lobule, but this view has gradually lost ground. Notwithstanding the statements just made that motor representation of voluntary movement is confined narrowly to the anterior central convolution, it must be admitted that there is an area anterior to this convolution, and particularly in the posterior end of the second frontal convolution, irritation of which produces movement of the eyes. Moreover, irritation of the posterior end of the first frontal convolution produces movement of the head to the opposite side.

**The Focal Symptoms of Disease of the Motor Cortex.**—The most striking symptoms which disease of the cortex of the brain produces are motor symptoms, which manifest themselves in two forms: In expression of irritation—twitchings, spasms, and convulsions; and of obliteration—paralysis more or less complete. As a rule, lesions that develop slowly, such as new-growths of various kinds, and lesions which are not of sufficient severity completely to overwhelm, such as poisonous matters circulating in the blood, produce irritative effects, while rapidly occurring lesions, such as hemorrhage, acute inflammation, and traumatic destruction, produce paralysis. In many instances, disease of the motor cortex is manifest first by irritative phenomena: twitchings and convulsions which, after a variable time—in one instance a few minutes and in another many months—are followed by paralysis. Occasionally paralysis follows immediately upon convulsions and is transitory in duration. In such cases the paralysis is interpreted as an acute exhaustion of capacity of the motor cells following an intense so-called motor discharge.

The distribution of spasm due to irritation of the motor cortex depends largely upon the area that is involved, although it does not absolutely correspond to this. An irritative lesion of the motor area of the brain may manifest itself in twitchings and spasms, limited to the corresponding contralateral peripheral parts, such as the phenomena known as Jacksonian epilepsy. It may, on the other hand, show itself first in these parts, but before the convulsive seizure has passed it may extend to the entire body. Spasm and convulsions caused by irritation of the motor cortex are usually followed by loss of consciousness, but in many instances, and especially in diseases that are of small extent and slight intensity, and those which develop very slowly, the twitching manifestations frequently occur many times (*i. e.*, over a protracted period) without accompanying loss of consciousness. Sooner or later unconsciousness becomes an accompaniment of the attack.

The antithesis of spasm is *paralysis*. When the motor cortex no longer responds to irritation, lesion of it is evidenced by paralysis. Paralysis, it matters not how brief its duration, indicates abolition of function of the motor cortex. The intensity and distribution of the paralysis depend upon the nature and extent of the lesion that produces it. A slowly growing tumor in the middle of the motor area may produce a slight, slowly increasing paralysis of one upper extremity, showing itself in a finger or



thumb, and gradually, as the tumor increases in size and encroaches upon other parts of the motor area, the paralysis involves the face and the lower extremity. On the other hand, a cortical hemorrhage following trauma may produce a complete hemiplegia within a few minutes. Circumscribed encephalitis may be so narrowly confined in the cortex that only the face and shoulder may be paralyzed for any considerable length of time, although the œdema, which is secondary to the inflammatory process, may cause a disturbance of motor function over a much more extensive area. As a rule, paralysis of cortical origin is characterized by incompleteness and by definite clinical phenomena: slowness of development, spasticity, diminution or abolition of the plantar-jerk, exaggeration of the tendon-jerks, these phenomena having been preceded or accompanied by spasm or convulsion, and particularly by the fact that it is confined in the beginning at least to one member of the body. Naturally, a lesion that obliterates suddenly the functions of the motor cortex causes paralysis of abrupt onset. In such cases spasm and convulsions are likely to be added to the clinical picture later when the results of the reparative process (scar tissue, adhesions) act as an irritant. The permanence of a paralysis depending upon a cortical lesion is intimately associated with the severity of the disease process. It is a common experience to have hemiplegia follow surgical interference of, or in the vicinity of, the motor cortex, which disappears after a few days or weeks.

**The Focal Symptoms of Disease of the Sensory Cortex.**—Knowledge of sensory localization in the cortex is much less definite than that of motor. The manifestations of lesion of the motor sphere are very striking, mainly objective, and not readily susceptible of misinterpretation. The manifestations of sensory irritation, on the other hand, are frequently indefinite, subject to wide variation, often confined to one variety of sensibility and not affecting another, and in every instance subject to personal interpretation.

For a long time it was believed that sensory representation in the cortex coincided very closely with motor, and this view, promulgated by Munk, is still held by many students of physiology, such as Horsley. It was thought that the Rolandic cortex was in reality sensorimotor or even kinesthetic (Bastian). Gradually, however, the advocates of the separate localization of the sensory and motor areas have gained ground, and the soundest teaching at present seems to be that the sensory area is confined largely to the posterior central convolution on the external surface of the brain, and to the falciform lobe on the mesal surface, particularly the central portion of the latter. On the external surface of the brain, sensory representation extends backward into the superior parietal convolution, to the anterior portion of which is allotted form-perception representation, while to the anterior part of the inferior parietal convolution is allotted muscular sensibility. Much effort has been made to give the various forms of common sensibility definite localization. The various sensibilities making up the "muscular sense" are thought to be located in the posterior half of the postcentral gyrus, the tactile, pain, and thermal senses in the anterior half of the same convolution.

Mills maintains that the cortical representation of cutaneous and

muscular sensibility is subdivided into a mosaic of centres, each of which is correlated anatomically and functionally to one or more motor centres, and that the areas of sensibility are subdivided like the motor area. Although this view is not yet accepted generally, there is much evidence in favor of it, quite enough to justify the position here taken.

**Allochiria.**—Allochiria (other hand) is the name given to a condition in which, though sensibility is retained more or less completely, the patient cannot tell which side of the body is being touched. The patient can feel the touch and can often describe the qualities of the object exciting it, but refers it to a spot on the opposite limb corresponding to that touched. E. Jones has also maintained that under the name of allochiria two fundamentally different conditions have been confused: (1) Part of a general defect in localization (alloesthesia), and (2) a specific defect independent of any error in localization (dyschiria). In other words, dyschiria of this writer, in reality, is allochiria of the majority of writers. Neither allochiria nor alloesthesia has any topical diagnostic value. All the evidence so far is in favor of the view that allochiria is a manifestation of hysteria, and does not occur with organic diseases.

**Astereognosis.**—Much clinical importance has been given to astereognosis and to the localization of the so-called "stereognostic sense." Great confusion exists because of the loose way in which the term has been used. Stereognosis means form perception, but most writers use it to mean the capacity to recognize objects by handling them, which incites a number of sense impressions. Burr contended that the "stereognostic sense" is not a sense but a judgment, and Prince emphasized this point. Unfortunately the term has come to have this application. In this sense stereognosis being an intellectual process, it cannot be localized any more than any other feature of the intellect. However, if it is limited to perception of form alone, tactual astereognosis may well have definite localization. The term agnosia, meaning inability to recognize objects because of lack of information about them, is a term that has a more legitimate usage. Tactual agnosia is the inability to recognize objects by touch. Such capacity may also have definite localization, inasmuch as the various forms of tactual sensibility may have definite localization, but no more. Disturbance of stereognostic perception may follow injury of the motor area, the ascending frontal convolution, and in a few instances it has accompanied disease of the area; but at the present time the best teaching is that astereognosis, associated with other symptoms of cortical involvement, indicates lesion of the superior parietal convolution, and especially its anterior portion in the vast majority of instances.

**"Soul Paralysis."**—We sometimes encounter cases of motor paralysis, that is, loss of voluntary and spontaneous motor action, in which there is no disease of the cortical motor areas or of their projections. Physiologists do not all admit that unilateral motor paralysis may result from destruction of the avenues that convey common sensation alone, but the preponderance of opinion is in the affirmative. It is certain, however, that when the cortical sensory centres or the tracts connecting them with the motor areas are destroyed, ability to perform voluntary

motor acts is lost and spontaneous motor power is impaired, occasionally wholly abolished. This condition, called "soul paralysis," "Seelenlähmung" of the Germans, was described by Munk to indicate a bizarre state noticed in dogs after extirpation of the sensory cortical area. Like other conditions or symptoms thought to be new, we find on examination of the older writers that it had not escaped their notice. There is no doubt that Duchenne described it under the name of "perd de la conscience musculaire." It is usually associated with profound sensory disturbances, and its most striking feature is due to a break in the sensory or afferent section of the reflex arc. Voluntary acts are really reflex acts and are possible only when the reflex arc is intact. An act performed in response to an effort of the will calls for a revival of the sensory memories that accompanied similar performances in the past and the stimuli that pass from them to the motor centre produce the movement that the will desires to produce. These afferent stimuli pass from the areas in which such memories are "stored." In practically all cases of soul paralysis that have been investigated anatomically there has been found destruction of the cortex or of the subcortical region of the parietal lobes or of the sensory tracts.

Soul paralysis is not always used in the sense here employed. Nothnagel used it to designate loss of the memory pictures of movement for one extremity or for one-half of the body. This condition Meynert called motor asymbolia. This word, and agnosia, apraxia, and other designations, which have been used with much latitude, are now coming to have very definite meanings attached to them. Their value in topical diagnosis has not been definitely assigned.

**Asymbolia.**—The word asymbolia was first used by Finkelnburg to denote inability to understand certain conventional signs aside from speech signs. Wernicke used it in this sense, but enlarged it to include failure to recognize the images of objects, *i. e.*, the loss of the "memory pictures" of the object. Thus, in Wernicke's sense, patients with asymbolia can see, hear, feel, smell, and taste, but they cannot re-recognize objects by these means. That is, they do not call up the previously existing memory pictures. They cannot turn sense impressions to account. The term asymbolia is therefore used by some in a sense not unlike that of agnosia.

**Agnosia.**—This is a term used to indicate inability to recognize things for what they are. An object is perceived by the peripheral sense apparatus, but it is not seen by the "mind's eye." Thus, there may be as many kinds of agnosia as there are special senses: visual agnosia (mind blindness), auditory agnosia (mind deafness), tactile agnosia, gustatory agnosia, and olfactory agnosia. Tactile agnosia is not the same as astereognosis, although it is frequently confounded with it. Before an object can be used properly, it must be recognized; therefore, agnosia entails dyspraxia, often apraxia, but apraxia does not necessarily indicate the existence of agnosia, in fact, there are many cases on record in which it existed without agnosia.

**Apraxia.**—This is the inability to perform purposeful familiar acts by a person who is not paralyzed, anesthetic, demented, or ataxic. It is a

commoner symptom of organic disease of the brain than is generally admitted. The fact that the localization of the lesion which causes apraxia is not so definite as that which causes other symptoms, spasm, for instance, explains why so little attention is given to it. The status of apraxia as a definite reliable localizing symptom is not yet satisfactorily established. Two varieties of it are distinguished—motor apraxia and ideational apraxia. In the first there is inability to translate the idea of a movement into a movement and in the second there is some defect in putting together the components of the idea of the movement. It generally involves both sides of the body, but it may be unilateral. Indeed, it may occur in one group of muscles only. Apraxia has been noted with cortical and subcortical lesions of various parts of the brain, but there is constantly increasing evidence to show that lesion of the anterior pole of the left hemisphere, especially the upper two frontal convolutions, will cause it, other portions of the brain cortex being normal. Lesions of the corpus callosum also cause it. v. Monakow is of the opinion that lesion of the gyrus supramarginalis causes apraxia.

Apraxia is associated with lesions of the left hemisphere more often than with lesions of the right. It has not yet been found with lesions below the capsule. The apraxia associated with disease of the left hemisphere may be homolateral or heterolateral. Although the lesions with which apraxia are commonly associated are those of arteriosclerosis and brain tumor it is not infrequent in the confusional states following attacks of epilepsy, in the dementia of alcoholism, and other toxic states. Like all symptoms, it may be caused by hysteria. Its most frequent symptomatic association is with aphasia.

**Amnesia.**—Although amnesia is the most constant symptom of disease of the brain, neither its intensity nor its variety has any relationship to topical diagnosis.

**Hemianopsia.**—The centre of sight (the primary cortical visual area, so-called) is more definitely located than any other. It is easily accessible in animals, and its blood supply is often disordered in man. Despite this, there is no unanimity of opinion on the part of the physiologist and the clinician regarding its limitations. The best opinion seems to be that it is situated in the mesial surface of the brain, around the calcarine fissure, in the cuneus of the occipital lobe. It occupies the cortex and the entire lingual lobe behind the junction of the calcarine and the parieto-occipital fissures, and the entire cuneus extending for half an inch or more on the external aspect of the occipital lobe. Destruction of this area in one hemisphere causes homonymous hemianopsia. It is now generally admitted that permanent visual disturbance occurs only when the entire visual area is destroyed. The primary cortical area for sight has two divisions, a peripheral and a central. The central is for the representation of the macula, and the peripheral for the remainder of the retina. The view of Henschen, that the centre for the macula is situated toward the apex of the cuneus, in the inferior extremity of the calcarine fissure, is the one that has received most substantiation. The lower quadrant of the field is represented in the upper half of the cuneus; the upper quadrant in the lower half of the cuneus.

The secondary, or higher visual area (visuopsychic of Campbell) is an association area variously allocated in the posterior end of the inferior parietal convolution, particularly the angular gyrus, or a considerable part of the parietotemporo-occipital cortex. Its function may be that of a storehouse of visual memories and a part of the mechanism for the appreciation of things seen.

The *symptoms* which disorder of the primary and secondary visual areas cause, which are available in topical diagnosis, are anopsia and hemianopsia, word blindness, and mind blindness. Cases of purely cortical anopsia and hemianopsia are extremely rare, for in the majority of such cases the optic radiations are found to be involved. Hemianopsia may be partial or complete, depending upon the extent of the primary visual area involved. It may also be double, as the result of second attacks of a bilateral homonymous hemianopsia.

In attempting to distinguish hemianopsia due to lesion of the cortex and to lesions of the optic radiations from hemianopsia dependent upon disease of the optic tracts and of the basal ganglia, the peculiar *pupillary reaction* described by Wernicke is of no importance. Wernicke pointed out that when hemianopsia was dependent upon involvement of the cortex and optic radiation, the pupillary response was intact for the entire retina, while when the optic nerves and optic tracts were the seat of the disease, only one half of the retina responded. This has never been corroborated by neurologists or ophthalmologists. All that may be said with positiveness concerning the attribution of hemianopsia to the cortex is that when it is not associated with other symptoms, such as sensory disturbances, pupillary disturbances, and aphasia, it is probably not of cortical origin. Further, quadrant anopsia is suggestive of cortical lesion.

Cortical color blindness, or achromatopsia, is not a symptom of importance in the diagnosis of focal disease of the brain. In other words, the distinction cannot be made clinically between color blindness due to disorder of different parts of the central ocular mechanism.

*Mind blindness* is the name given to the condition in which an affected individual is unable to recognize familiar objects by the sense of sight. In other words, it is a condition in which visual memories of things previously seen and comprehended cannot be reinvoked. Mind blindness is dependent upon disease of the cerebral cortex, or possibly of tracts which assist certain parts of the cortex. The term is here used to indicate object blindness, word blindness, letter blindness, and symbol blindness, with intactness of the peripheral ocular apparatus and the primary visual centre, and due to lesion or disturbance in function of the higher or secondary visual area, and possibly also to lesion of the tracts or association fibres which connect the higher visual field with the lower visual centre.

**The Centres of Hearing and Smell.**—The centre for hearing is situated in the posterior transverse temporal gyri and the cortex of the first and possibly the second temporal convolutions. By virtue of the hemidecussation of the secondary auditory fibres in the pons each centre has a bilateral representation. Permanent deafness would therefore not be expected from a one-sided cortical lesion.

*Auditory disorder*, aside from word deafness, is not an important symptom suggestive of topical diagnosis. Subjective auditory symptoms, auditory hyperesthesia, and variously described paresthesia may occur with tumor or other slowly developing lesion of the pons, or at the base of the brain encroaching upon the auditory nerve. Similar symptoms preceding an epileptic attack should suggest lesion of the temporosphenoidal lobe of the opposite side. Deafness of some degree is the result of disease of the central auditory apparatus. When it is of the cortex, it is called word deafness (see aphasia); when of other part of the central auditory mechanism, nerve deafness. Instances of bilateral deafness from tumors of the quadrigemina encroaching upon the tegmentum have been recorded.

The centre for *smell* is situated in the uncinate gyrus of the same side, and probably to a certain extent on the opposite side. Jackson and other clinicians have shown that this gyrus is diseased in cases of epilepsy attended with olfactory auræ, such as unaccountable smells. Disorder of the sense of smell, especially when associated with symptoms indicative of brain lesion, should suggest at least an involvement of the uncinate gyrus, but it has also existed with tumor of the cerebellum.

The higher mental faculties, such as abstraction, introspection, judgment, comparison, reasoning, etc., are subserved mainly by the brain anterior to the motor areas, particularly in conjunction with association areas in other parts of the brain. So far nothing definite has been established concerning the subdivision of the prefrontal lobe into centres to which are allocated definite components of the higher intellectual functions, and, considering how complex the higher psychical faculties are, and that modern psychology views all mental faculties as arising from a complicated coördination or adjustment of movement to the environment, it is perhaps not legitimate to seek to localize them.

Many investigators believe that the anterior part of the frontal lobe probably is concerned in selecting and coördinating the products of associative process of the post-Rolandic area and converting them into psychomotor manifestations or inhibiting them, while the posterior is purely a psychomotor area in which physical complexes necessary to give external expression to the results of cerebral association are developed and elaborated.

There is much convincing evidence to show that Flechsig's anterior centre of association is more concerned with attention and other coördinations of psychic process than any other part of the brain. It is an area undeveloped in cases of idiocy and diseased in cases of secondary dementia. Although there is no centre of attention or thought or apperception or morality in the frontal lobes or elsewhere in the brain, the frontal lobes contain neural elements whose integrity must be maintained if the individual is to develop and persist in habits of attention, concentration of thought, balance of feeling, sound judgment, and moral conduct.

Lesion of the frontal lobes, aside from the so-called ascending frontal (which in this article is referred to as the anterior central), and the foot of the third frontal convolution (Broca's convolution), produces no constant, definite, localizing symptoms. In the majority of cases, however, such

lesion is attended with distinct mental symptoms, such as inattentiveness, lack of concentration, impaired judgment, emotionality, and impairment of the higher mental faculties, reaching even to imbecility. If the lesion is of the orbital surface of the frontal lobe, olfactory and optic tract symptoms often occur even in the early stages of the disease. Such symptoms followed by motor symptoms, disturbance of speech, monoplegia, and hemiplegia, are highly suggestive of lesion in this part of the brain.

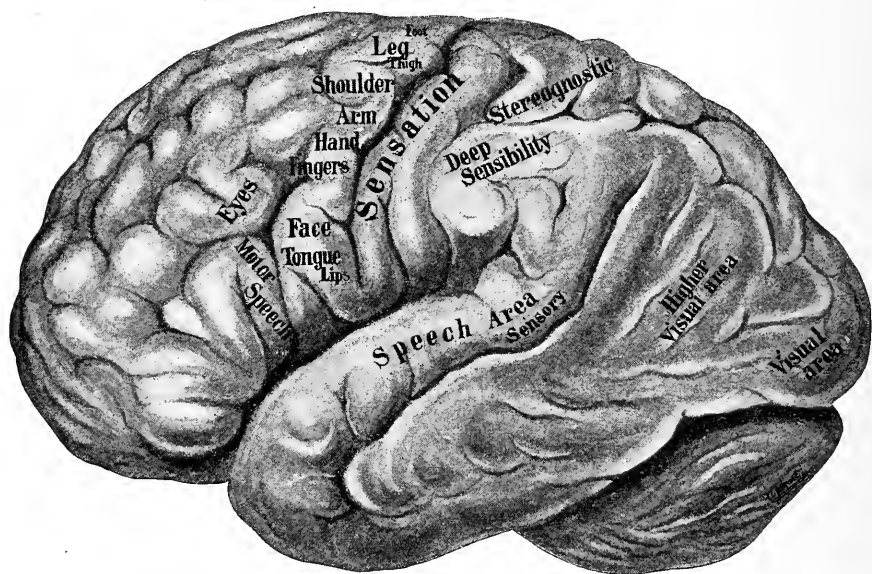
Grainger Stewart suggests that absence, diminution, or easy exhaustion of the superficial abdominal reflexes on the contralateral side and a fine, rapid, vibratory tremor in the limbs of the same side are suggestive of tumor of the frontal region of the brain, but I have seen these symptoms in cyst of the post-Rolandic area.

**Summary.**—Briefly to summarize the present-day teachings of localization of function in the cortex: The motor area and the sensory area are situated in the centroparietal region, the motor anterior to the fissure of Rolando, the sensory posterior. These areas are subdivided so that different portions of them serve different muscles and different varieties of sensation. An inverted manikin, parallel to the fissure of Rolando, will serve to remind one of the divisions of the motor area. The centres for cutaneous and muscular sensibility are in the posterior central convolution; that for so-called stereognostic sensibility in the anterior end of the superior parietal and probably the adjacent posterior central. The centre for vision is in the occipital lobe, particularly in the cortex around the calcarine fissure, that for hearing in the middle third of the first temporal lobe, and that for smell in the hippocampal region. The left hemisphere contains the speech area in right-handed individuals, and *vice versa*; the part of the brain that subserves this function includes the foot of the third frontal, the superior temporal, and the posterior end of the inferior parietal convolutions. In addition to these centres, the cerebral cortex has special constituents serving for association processes, which are located by some in definite centres, but which are by the majority assumed to be scattered over the entire cortex. Speech is related both to the sensory areas and the associative areas. On the one hand, the word-component of ideation is dependent upon the presence of association fibres; on the other hand, it is based upon various sensations which have their seat in the sensory centres. Finally, the frontal lobes are necessary for the maintenance of those processes which underlie the higher psychological faculties.

If these areas have the functions that have been allotted them, disease of these areas should cause definite symptoms, and it is a fact that it does. It would seem, therefore, that topical diagnosis of brain disease should not be difficult, and it would not be were it not that (1) the disease process is not often confined narrowly to one of these areas; (2) the immediate and subsequent result of disease of these areas is disturbance of function of other areas both proximate and remote; (3) disease of one area may be but part of a widespread disease of the vascular system, the centre that apparently manifests the disease being either the one which is bearing the brunt of the lesion or is most tolerant of encroachment; and (4) in every case of topical disease of the brain there are general symptoms which

vary with the nature, seat, extent, and duration of the lesion. Such symptoms occur practically in every case, and are called the general symptoms. They are headache, dizziness, vomiting, convulsions, apathy, stupor, disturbance of gait and station, disorder of intellect usually manifest by sluggishness of the so-called psychical reflex, and disorder of the optic nerves. This, in conjunction with the fact that a considerable extent of the brain cortex and almost all of the brain substance beneath the cortex has no definite functional allotment, makes accurate topical diagnosis difficult. Occasionally when a lesion is confined narrowly to one of the areas whose function has been definitely determined, especially when the disease process is small and of slow onset, so as not to cause obscuring symptoms, the diagnosis may be made with something approximating mathematical accuracy.

FIG. 18



Localization of function in the brain cortex.

In every case the most essential thing for such diagnosis is careful, repeated physical examination, the results of which are noted and, after a sufficient number of them have been made, compared. If, then, there are persistent constant disturbances of function of one or more of the centres, which have special function, and likewise one or all of the general symptoms, a definite topical diagnosis is possible.

**Lesion of the Central Convolution.**—Lesion of the anterior central convolution causes spasm of a muscle or of functionally associated groups of muscles, such as those of the leg, the hand, and the face, of the opposite side, or paralysis, partial or complete, in similar parts. Such paralysis is nearly always spastic unless the lesion is traumatic and dependent upon extensive laceration or hemorrhage, when it may be flaccid. If



the lesion extends backward into the posterior central convolution and the adjacent parietal convolutions, disturbance of muscular, cutaneous, postural sensibility and impaired recognition of the forms and uses of objects, so-called astereognosis, will be present. For instance, the occurrence of a series of tonic or clonic spasms in the hand, followed or not by loss of consciousness and other manifestations of an epileptic attack, would immediately suggest lesion of the middle third of the motor area. In case such symptoms are followed by slowly developing monoplegia, and later by such sensory disturbances as impairment of the deep sensibility and astereognosis, the justifiable assumption is that the lesion starting in this area had extended posteriorly.

A lesion confined narrowly to the posterior central convolution does not produce pure motor phenomena, but causes impairment of tactile sensibility and of muscular and articular sensibility, while lesion of the anterior end or the superior parietal convolution, possibly also of the posterior central convolution, produces tactile astereognosis, tactile asymbolia, hemiataxia, and abolition of the superficial reflexes, especially the plantar reflex, and in some instances the abdominal. Extension of the lesion from this area into the motor area is accompanied by motor manifestations and exaggeration of the tendon-jerks, while extension backward to involve the cuneus or in depth to implicate the fibres of Gratiolet is followed by hemianopsia. Lesion of this area is usually not attended with psychical symptoms or disturbance of speech until late in the disease.

Lesion of the occipital area, and particularly of the cuneus, produces homonymous hemianopsia, which is often associated with word blindness in conjunction with the general symptoms of intracranial disease. This is often a most important localizing symptom. Visual hallucinations, transitory but recurring, are occasionally early symptoms of disease of the occipital lobe. Such are referred to one-half of the visual fields.

**Lesion of the Centrum Semiovale.**—The white substance of the brain is made up largely of fibres that convey impulses to and from the cortex and fibres that connect different parts of the same hemisphere or one hemisphere with the other. Disease of it should therefore be evidenced by symptoms varying with the variety and number of the fibres involved, *i. e.*, varying with the situation and size of the lesion. If the fibres coming down from the motor centres to pass through the internal capsule to the pyramidal tract are involved, and these only, the prominent symptom will be paralysis of the opposite side of the body. Lesion of this sort, unless close to the cortex, is not so frequently attended with convulsions, particularly of the localized variety, as is lesion of the motor cortex itself. On the other hand, general epileptoid attacks often occur. If, again, the lesion interferes with or interrupts the sensory fibres radiating to the posterior central and superior parietal convolutions the symptoms will be predominantly disturbance of sensation with hemiataxia. If the lesion is in the posterior part of the white substance, then symptoms referable to vision or to the speech faculty are likely to predominate. A lesion of the white substance of the brain in the anterior pole cannot ordinarily be diagnosed or even suspected. As a rule, the general symptoms are those of intracranial disease with no localizing symptoms. As

a matter of fact, lesion of the centrum semiovale, such as tumor, often gives rise to symptoms thought to be characteristic of remote parts of the brain, as the cerebellum.

**Lesion of the Corpus Callosum.**—Disease of the corpus callosum, particularly tumor, causes symptoms which, although not pathognomonic, are suggestive of invasion of this portion of the nervous system. Mental symptoms occurring early are most characteristic. These are peculiar delusional states, eccentricity of manner and action, impairment of memory, dulness, excitability, want of sequence in ideas coupled with an apparent conservation of the intelligence, followed by the general symptoms of intracranial disease, which may, however, develop very late. Apraxia has been recorded and irritative and paralytic phenomena due to encroachment upon the neighboring motor zones. An attempt has been made at a subdivision of localization in the corpus callosum, but this is an unnecessary refinement of diagnosis.

**Lesion of the Internal Capsule.**—This is the avenue which impulses originating within the brain must traverse in order to be externalized, and likewise the avenue through which impressions coming from without must pass in order to be interpreted by the brain. In other words, it contains practically all the projection fibres of the cerebral cortex. Therefore, lesion of the internal capsule may produce symptoms that parallel those of lesion of the cortex. A small lesion in this area of the brain will be accompanied by symptoms which could otherwise be caused only by most extensive disease of the brain cortex. Complete destruction of the internal capsule produces flaccid hemiplegia, hemianesthesia, vasomotor disturbances of the paralyzed side of the body, hemianopsia, and disturbance of hearing on one side.

The internal capsule, which is in the form of an obtuse angle, is divided by the apex of the angle, or the knee, into two portions, an anterior and a posterior. The anterior part, or the lenticulo-caudate segment, contains the fibres from the frontal lobes, particularly those going to the thalamus. The posterior division, the lenticulo-optic and the retrolenticular segments, contains, in its anterior part, the pathway of the lower third of the precentral convolution. These fibres are located in the anterior part of the posterior division. They pass from there into the ventral half of the pons, to end mainly around the nuclei of the facial, the motor nucleus of the trigeminal and hypoglossal nerves. A lesion of this portion of the internal capsule, it is maintained by many, especially by the school of Charcot, causes symptoms referable to these nerves, particularly paralysis of the muscles of the face which are under volitional control. The pathway made up of projection fibres coming from the other regions of the motor cortex, and known as the pyramidal tract, especially after it reaches the pons, is situated just posterior to the tract mentioned above. These two pathways or fascicles constitute the anterior two-thirds of the posterior limb. Adjacent to this, posteriorly, is the pathway of the general sensibility from the entire body, which, coming in from the tegmentum of the crus, passes up through the internal capsule to go to the parietal convolutions. Capsular hemianesthesia of organic origin may be accompanied by hemianopsia, but not sensorial hemianesthesia.

When alteration of the most posterior segment of the internal capsule is attended with hemiplegia accompanied by hemianesthesia and hemianopsia, the optic tract participates in the capsular lesion. Between the pyramidal tract and this fascicle, practically a portion of the former, Charcot, Raymond, and others place the "fascicle of hemichorea," *i. e.*, the portion of the posterior limb of the internal capsule, irritation of which causes hemiathetosis, unilateral paralysis agitans, unilateral intention tremor, and hemiataxia. Finally, the visual tract passing from the pulvinar of the thalamus to the occipital lobe, and the auditory tract going from the auditory nucleus to the temporal lobe, complete the internal capsule.

This being the relative composition, functionally, of the internal capsule, it follows that lesion of different parts of it produces different symptoms. Clinically there is very little satisfactory evidence that such localization exists. Horsley and Beever demonstrated, apparently to the satisfaction of physiologists, that such localization exists in the bonnet monkey, and in most of the recent standard works on pathological anatomy and neurology it is stated that it is true for man. But Marie maintains he is not able to corroborate such teaching and the writer's experience leads him to agree with this. The teaching that there is definite capsular localization has been largely the outcome of the acceptance of statements of Charcot and his school; and of homologizing in man the results of experiments on the lower animals. It is impossible to say that there is a special territory of the internal capsule of man that is exclusively sensory, and even with intense capsular and cortical lesion, hemianesthesia may be absent.

Marie maintains that the motor pathway occupies the entire posterior segment of the internal capsule as far as the level of the posterior angle of the lenticular nucleus. In this lenticulo-optic territory, in which descend motor fibres, it is impossible from clinical data to specify distinct territories for the fascicles that go to the arm, leg, foot, etc. A lesion of this part of the capsule produces the syndrome of hemiplegia, and that is all, and, conversely, a lesion limited to the internal capsule causes degeneration of the entire pyramidal tract at the level of the oblongata or the cord. Clinically one cannot establish any persistent segmentary localization in the internal capsule. Very small lesions of the posterior limb of the internal capsule may produce irritative symptoms manifest as contracture and irregular movements on purposeful effort. After a considerable vascular lesion has been partly absorbed, the destruction of the pathway will be manifest by hemiplegia and the irritative effect on the partially destroyed fibres may be evidenced by contracture. When the lenticulo-optic section of the internal capsule is diseased, there may be a flaccid, complete hemiplegia with, later, hemicontacture. Conjugate deviation of the eyes associated with forced movements of the head is sometimes an initial symptom. If the part of the internal capsule just anterior to the knee is affected, there will be hemiplegia and dysarthria, or, if it is a small area in front of the knee, facial paralysis alone may be the symptom.

Occasionally a focus in the lenticular nucleus or in the optic thalamus

encroaches upon the internal capsule in such a way as to produce a characteristic clinical picture, paralysis of the face and of the leg, while the arm is spared. In such cases the lenticulostriate and the retrolenticular section of the internal capsule are affected, while the lenticulo-optic section remains comparatively free.

Double-sided lesion in the internal capsule, like certain bilateral cortical lesions, produces the condition known as pseudo-bulbar paralysis, and, varying with the encroachment upon the pyramidal tract, hemiplegia of one or both sides.

**Lesion of the Corpora Striata (Ganglia of the Cerebral Hemispheres); the Lenticular Nucleus and the Lenticular Zone.**—The corpora striata, the greater part of which is embedded in the white substance of the hemisphere, are constituted of two parts, an intraventricular part, called the caudate nucleus, and an extraventricular, called the lenticular nucleus. The functions of this body have not been satisfactorily established. Dejerine denies that it has any connection with the motor and other portions of the cortex or that it plays a part in the control of movement or in the function of speech. Mingazzini, on the other hand, has proved conclusively that certain parts of the lenticula transmit motor and sensory fibres and have motor and sensory functions. A focus of disease, even of small size, involving only the lenticular nucleus, never fails to cause motor disorder, usually partial paralysis, to which, sometimes, are added irritative symptoms. Motor paresis of the entire half of the body, often of slight intensity and therefore overlooked, is the most frequent symptom. This often disappears, but a residue can always be found by careful examination. Disturbance of speech is also often associated with lesions of the left lenticula. The lesion in the majority of cases with the syndrome of hemiparesis has been softening of the putamen. The motor functions of the lenticula are supplementary to the pyramidal tract. There is no justification for regarding it as a vestigial organ; Mills and Spiller have furnished ample proof that destructive lesions of certain portions of the lenticula cause paresis of the limbs or face, which differs from that produced by capsular lesions in so much as the impairment of power is not so severe, and from cortical lesions in that it is less likely to be dissociated, and Kinnier Wilson<sup>1</sup> has described a well-defined clinical picture of Progressive Lenticular Degeneration, due to bilateral progressive diseases of the putamen in particular.

**Progressive Lenticular Degeneration.**—Clinically, this disease manifests itself in involuntary movements: tremor-dysarthria, dysphagia, muscular weakness, spasticity and contractures with progressive emaciation, usually associated with emotionalism and often with mental symptoms. It is a disease occurring in young people, often familial, but not congenital or hereditary. The course is progressive and the termination fatal. Pathologically, it is characterized by a bilateral symmetrical softening of the lenticular nucleus, especially of the putamen, while the globus pallidus is less affected. In addition, *cirrhosis of the liver* is constantly present. In pure cases the internal capsule is absolutely intact. In

<sup>1</sup> *Brain*, 1912, xxxiv, 295.

other words the affection uncomplicated represents an extrapyramidal motor disease.

Marie maintains that the lenticle and the area around it, which he calls the lenticular zone, play an important rôle in the causation of motor aphasia. There is no doubt that lesion of this zone causes disturbance of speech, the result of impairment of coördination of the complex muscular movements concerned in the exteriorizing of speech, and apparent motor aphasia must be considered a symptom of disease of the lenticular nucleus.

Certain symptoms, mental and physical, accompany with much constancy disease of the globus pallidus (the inner part of the lenticular nucleus), due to illuminating gas, to which the lenticle is peculiarly vulnerable. These are vasomotor and trophic disturbances of any of the soft parts (discoloration of the skin, ulceration simulating bedsores, etc.), disturbed control of the bladder, very rarely of the bowels, amnesia or loss of memory, especially the variety known as retrograde, and a peculiar mental state, which shows itself most strikingly in the appearance of the patient, reminding one of catatonia. Such patients are fully oriented, apparently appreciative of all that goes on around them, but wholly indifferent, apparently uninterested, reflecting in their features and attitudes absence of initiative.

**Lesion of the Optic Thalamus.**—It has been taught with much confidence that the optic thalamus is a region which manifests disease or encroachment upon it by motor, sensory, secretory, and emotional symptoms. These are hemiplegia, posthemiplegic choreic movements, intention tremor, hemiataxia, disorder of involuntary mimetic movements, hemianesthesia, hemianopsia, vasomotor and trophic manifestations in the contralateral extremities and pain.

The "*thalamic syndrome*" has been rather widely accepted. Gradually, however, it has become obvious that there are two reasons why it should be subject to critical investigation: First, because destruction of parts of the optic thalamus by disease may occur without symptoms; and second, because many of the cases upon which the claim has been made that lesions of the optic thalamus caused such symptoms have been investigated only macroscopically, and therefore it is impossible to say that the subthalamus, the red nucleus, and even the retrolenticular constituent of the internal capsule were not affected as well. Finally, many cases of thalamus lesion which did not produce the above-mentioned symptoms have been carefully studied.

Lesions confined to the thalamus do not cause hemiplegia. Although hemiplegia occasionally occurs as an initial and transitory accompaniment of thalamus lesions, its existence is to be interpreted as a coincidence, or as the result of a lesion at a distance. In other instances in which hemiplegia has been present with lesion of the thalamus, careful examination of the crura (cerebral peduncles) or the capsule has not been made. Certain other disturbances of motility occur in thalamic lesions, especially lesions of vascular origin. These are so-called post-hemiplegic choreic movements, intention tremor, ataxia of purposeful movements, athetoid movements, and disturbance of the involuntary

mimetic musculature. Lesion of the pulvinar and of the lateral geniculate body causes spasticity of the contralateral extremities and ataxia or tremor especially of the upper extremity. In some instances the movement which accompanies such lesion is more or less deliberate and rhythmical, known as athetoid. The term athetosis, meaning without fixed position, was introduced by Hammond to describe slow, apparently determinate systematic movement with a tendency to distortion of an extremity or part of an extremity, such as the hand or fingers. Athetosis is probably always the result of thalamic lesion and not of lesion of the corpus striatum. Choreic movements, ataxia, and intention tremor may result from lesion in the cortical motor pathways, but athetosis does not. In several cases of athetosis lesions of the red nucleus, the tegmentum, and the vicinity have been found.

Disturbed innervation of the muscles that cause involuntary emotion display is perhaps the most constant symptom of lesion of the thalamus. It has long been established by clinical observation that there are patients with slight hemiplegia or hemispasticity and other symptoms indicative of thalamus lesion, such as paresthesia of one-half of the body, disordered muscle sensibility, choreic or athetoid movements, who have the peculiarity that they cannot innervate the emotional muscles of the face voluntarily, but they are able to smile and weep emotionally. Many of these cases have been carefully studied, and lesion of the thalamus is invariably found. This symptom constitutes an important feature of the thalamic syndrome. The occurrence of forced smiling and weeping is due to lesion of the corticothalamic fibres, the mimetic centre reacting convulsively to every stimulus whether from the periphery or from the cortex. It should be said that Roussy claims the mimetic capacity was not disordered in the animals in which he produced lesion of these fibres.

The occurrence of *sensory* symptoms with lesion of the thalamus is by no means constant, although it would seem to be conclusively proved that sensory disturbances may be caused by experimental lesion of the thalamus in the lower animals. Moreover, there are many cases of thalamic disease with sensory disturbances, but in these cases there has not been adequate investigation of the state of the surrounding parts, only possible by microscopic examination. For instance, Dejerine and Long are inclined to interpret the sensory disturbances attending thalamic lesion as evidences of co-existing lesion of the retrolenticular division of the internal capsule. Lesions of the basal portion and of the subthalamic region are undoubtedly attended with sensory disturbances more constantly than lesion of the thalamus elsewhere. The disorder of sensation is usually limited to certain qualities, such as the tactile. Pain and temperature sensibility are sometimes undisturbed. Another feature of the resulting tactile and muscle anesthesia is that they are not permanent. This feature explains in a measure some of the cases recorded in which lesion of the thalamus was not accompanied by sensory disturbances, and especially cases of tumor which are of slow growth and long duration. In such cases there may have been sensory disturbances.

Vasomotor and trophic disturbances which occur with thalamic lesion are manifest particularly in the contralateral extremities. They are sub-

jective and objective sensations of warmth or of cold, redness, swelling, and hyperidrosis. Such symptoms have been observed so often with thalamic lesions that many writers maintain the existence of vasomotor centres in the thalamus. Numerous cases have been reported of hemiplegia with sensibility disturbances on the paralyzed side, in which there has been well-marked muscular atrophy on the opposite side, a condition that cannot result from lesion of the pyramidal tract alone. Some interpret this atrophy as the manifestation of a trophic influence, others as a vasomotor disturbance.

In brief, it may be said that the *thalamic syndrome* consists of (1) a persistent partial hemianesthesia, (2) slight hemiplegia, (3) ataxia and astereognosis of the contralateral half of the body, (4) pains, and (5) choreic and athetoid movements on the hemiplegic side. Head has called attention to another symptom, namely, an over-response to affective stimuli which he believes pathognomonic of thalamic affection.

Our knowledge of the symptomatology of thalamus lesions is indefinite because lesions limited closely to the thalamus are of rare occurrence, save tumors, and of such slow development and protracted course that compensation occurring meanwhile makes it difficult to say that the symptoms are, indeed, due to thalamic disease. Vascular lesions are rarely confined to the thalamus, and it is not at all improbable that many symptoms produced by them are due to simultaneous disease of parts of the brain adjacent to the cortex. von Monakow maintains that the thalamus constitutes an important station within the central nervous apparatus for sensory and cutaneous perception. The representation of individual sensory organs is present in it in a sharply localized way. The final relation of the anatomical connections must be deemed bilateral. Further than this, it must be accepted that other parts of the brain may functionate vicariously for the thalamus. In this way is to be explained the fact that symptoms of focal disease of the thalamus are transitory and that many instances of lesion of this region run their course without symptoms.

**Lesion of the Crura Cerebri and Tegmentum.**—After the motor fibres pass through the internal capsule they leave the cerebral hemisphere by the crus, of which they occupy the middle three-fifths. In their downward course they supply contralateral fibres to the nuclei of the motor cranial nerves. Lesion of a crus may be limited to the crura or to the tegmentum. When confined to the crus, it causes a characteristic syndrome, paralysis of one-half of the body on the side opposite the lesion, and of one or more of the cranial nerves, usually the third, on the same side, the so-called paralysis alternans.

**Tegmental Syndrome.**—The tegmentum of the crus cerebri, which is made up of a continuation of the formatio reticularis of the oblongata and pons, the superior peduncle of the cerebellum, and other white and gray matter, is sometimes the seat of disease which confines itself rather narrowly to it. Such disease produces fairly constant symptoms, which are said to constitute the "syndrome of the tegmentum." These are alternating sensorimotor hemiplegia, paralysis of the associated movements of elevation and depression of the eyes, of convergence, and

disorder of the internal musculature of the eyes with preservation of the associated movements of bilaterality. Such symptoms may be associated with involuntary movements (in rest and in action), hemi-ataxia, adiadokokinesia, hemihypoesthesia, and abolition of deep sensibility of the opposite side of the body.

Gruner and Bertolotti described two typical cases of this affection. The sensory hemiplegia was more marked than the motor, but disorder of involuntary movement, incoördination and asynergia, which superimposed themselves upon the hemiplegia, were well marked. To the hemiplegia was added bilateral and symmetrical paralysis of all the muscles innervated by the third and fourth pairs of cranial nerves. The only conjugate movements preserved were those of laterality. The lesion was a tubercle situated in the middle of the tegmentum at the level of the nucleus of the third nerve. It was bordered on its left side by a zone of softening which reached through the median lemniscus and totally destroyed the anterior quadrigeminal body. It had caused complete destruction of the gray column of the motor nuclei of the aqueduct extending from the level of the third ventricle into the pons as far as the level of the nucleus of the superior oblique.

**Lesion of the Quadrigeminal Bodies; Mid-brain.**—Disease confined closely to the quadrigeminal bodies causes symptoms from which a localizing diagnosis may be made with some confidence, but such disease is comparatively rare, especially hemorrhage and softening, which are of common occurrence in other parts of the mid-brain and hind-brain, but rarely confined to these bodies alone. Tumor, however, is sometimes so confined; it is from consideration of the symptoms accompanying such lesion, and from experimental work, that the symptomatology of disease of this part of the mid-brain has been made out. Experiments on the lower animals seem to show that destruction of the entire roof of the corpora quadrigemina produces complete blindness, and that lesion of one-half of it produces hemianopsia, but in man and the higher animals this does not occur, although in man an important optic nerve bundle is in connection with the anterior corpora quadrigemina. Disturbances of vision, even complete blindness, occurring in affections of the corpora quadrigemina have been described by many authors, but the fact that there are a great number of negative cases and cases in which there is no disturbance of vision during the entire course makes it certain that other parts of the mid-brain than the quadrigeminal bodies were implicated in those instances in which vision was affected. Profound visual disturbance accompanying such lesions is indicative of associated lesion in the external geniculate bodies or in the optic tract. Ferrier and Turner concluded from their experiments on monkeys that the quadrigeminal bodies have only a subsidiary relation to the special senses of sight and hearing, although it is generally believed that lesion of the posterior quadrigeminal bodies causes defect or abolition of the sense of hearing. They are also of the opinion that the sensory auditory tract lies outside the quadrigeminal bodies, the posterior quadrigeminal body being merely an accessory structure and end-station of the fibres of the lateral fillet; clinically, disturbance of hearing is not noted with sufficient constancy



to justify us in placing much reliance upon it as a localizing symptom of disease of the posterior quadrigeminal bodies.

Much attempt has been made to distinguish between disease of the anterior and the posterior quadrigeminal bodies. A lesion confined to the anterior pair causes bilateral paralysis of all the eye muscles save the abducens, while if the posterior are affected alone, ocular palsy is not an important symptom, but deafness and ataxia are.

Foci of disease of the quadrigeminal bodies adjacent to the tegmentum or the crura cannot be differentiated from lesions of these structures, but foci in the roof of the quadrigeminal body produce symptoms which are more or less characteristic. These are disturbance of vision, pupillary disturbances, limitation of movements of the eyeball, *i. e.*, some degree of ophthalmoplegia, especially of the oculomotor and trochlear nerves, sensory disturbances, and disorder of voluntary motion and gait. These symptoms vary with the extent of the lesion and with the implication of the surrounding bodies. This fact cannot be too strongly emphasized.

According to Bach, double-sided complete destruction of these bodies, but more particularly of the roof, causes lesion of the pupillary light reflex on both sides, while unilateral destruction causes it only on one side. But from experiments on the higher animals it seems to have been conclusively proved that there is retention of iridomotor action after complete ablation of the quadrigeminal bodies. This seems to indicate that the pupillary fibres of the optic nerve reach the third cranial nucleus by some other route than through the anterior quadrigeminal bodies.

The truth of the matter is that until a lesion of the corpora quadrigemina extends basalward, involving the tegmentum or the central gray matter around the aqueduct of Sylvius, the location of the lesion can only be suspected, but when it does so extend, fairly characteristic symptoms occur, the most important of which is ophthalmoplegia. A lesion confined narrowly to the corpora quadrigemina, and causing no pressure around the surrounding parts, is not accompanied by paralysis of associated ocular movements. The many cases in which the corpora quadrigemina have been destroyed without causing disturbance in the movements of the eyeballs attest this statement. Complete ophthalmoplegia is rare, but partial limitation of the associated movements of the eyes, especially the upward and downward movements, is fairly common, as is also ptosis. Ophthalmoplegia in connection with ataxia resembling cerebellar ataxia constitutes a most important association of symptoms. When the ataxia develops first and is very severe, the lesion is very likely to be in the cerebellum; when ocular palsies develop first, the ataxia much later, the lesion is likely to be in the corpora quadrigemina. Limitation of the ophthalmoplegia to the oculomotor and the trochlear, speaks in favor of affection of the corpora quadrigemina. Involvement of the abducens may occur both in quadrigeminal and cerebellar lesions, but involvement of other cranial nerves from the abducens downward indicates an affection of the cerebellum.

Paralysis of the trochlear nerve on one or both sides and disturbance of the chewing capacity have been described as symptoms of mid-brain disease, and especially of the posterior quadrigeminal bodies, the reticular

formation and the locus cœruleus. Such paralysis is manifest by difficulty of moving the eyes outward and downward and circularly. In the majority of these instances the paralysis is associated with hemi-ataxia, cerebellar ataxia, choreic movements, and oculomotor palsy. Its occurrence bespeaks extensive involvement of the central gray matter and of the thalamic region. Difficulty in chewing, pointing to involvement of the descending root of the fifth nerve, in reality a pons symptom, has been mentioned in a number of cases of quadrigeminal disease. Some writers speak of an association of symptoms consisting of atrophy in the region of the neck and shoulder, and later in the muscles of mastication, as the "Syndrome of Kojevnikoff," and maintain that it is dependent upon lesion of the posterior quadrigeminal bodies.

Disease of the mid-brain involving the tegmentum, particularly a vascular lesion, disorder of the functions of the red nucleus, the fillet, the reticular formation, and other pontile connections, is always accompanied by ataxia of a cerebellar nature often confined to one side of the body. In fact, this symptom, in conjunction with irregular choreic movements and intention tremor occurring on the opposite side of the body, is perhaps the most characteristic combination of symptoms of mid-brain disease. The ordinary ataxia of movement is unquestionably dependent upon involvement of the fillet and of the reticular formation. The "cerebellar" ataxia is not so easily explained and no satisfactory explanation has been given.

A lesion, such as glioma, affecting the gray matter around the aqueduct of Sylvius at a point midway between the two corpora quadrigemina and the third ventricle, may produce forced movement with a constant tendency to fall backward or to bear backward against a support, loss of the upward movements of the eyeballs and upper eyelids, exaggeration of the knee-jerks, ankle clonus, patellar clonus, and general symptoms of brain tumor.

**Lesion of the Cerebellum.**—The symptoms of disease of the cerebellum as a whole are often pathognomonic. The symptoms of disease of definite portions of it are equivocal. Symptoms of disease of the cerebellum vary profoundly with the suddenness of onset and the rapidity of progress of the lesion, and without full recognition of this fact the diagnostician is bound to be disappointed frequently. As Hughlings Jackson wisely said, "the symptoms are so different in different cases that it is nearly impossible to make general statements."

Some of the chief functions of the cerebellum have long been established, but it is only recently that we know with any certainty that there is definite localization of function in it. The cortex is a massive recipient organ (Flourens, Jackson, Edinger, Horsley), the head ganglion of the proprioceptive system (Sherrington), to which different impulses from all parts of the body converge for rearrangement and coördination, so as to render them suitable for transmission and interpretation to the deep nuclei, which may be looked upon as stations interposed between the different cortical fibres and the rest of the nervous system. The relation of these nuclei to the cortex has been fairly well established by experimentation, but the relation of the nuclei to the extracerebellar

nervous system is only now being determined. The chief rôle of the cerebellum is to collect and arrange the various afferent impulses outside consciousness for other parts of the nervous system. This is manifested in motor coördination, equilibration, and in the maintenance of muscular tonus. The cerebellum is also intercalated into sensory pathways which receive excitation from the periphery and from the higher centres, but its functional relationship to the sensory sphere becomes more doubtful the more experimentation forces the secrets of its purpose. The middle lobe is functionally more important than the lateral hemispheres, and removal of half of this lobe in lower animals causes disturbances similar to that caused by removal of a lateral lobe. The effects of unilateral ablation are manifest in disturbance of equilibrium, peculiar attitude, disordered sensibility, and ocular symptoms. In human beings injury and disease of the cerebellum cause similar symptoms and also disorder of speech.

The results of experimental lesion and destruction of portions of the cerebellum are very soon compensated by the sensorimotor cortex, and after a few months animals that have been thus operated upon present no abnormalities unless very careful examination is made. If the Rolandic area of the brain, however, is destroyed, simultaneous compensation does not result, and the cerebellar symptoms persist. In the human being injury to the cerebellum early in life, and arrested development of the cerebellum, may not give rise to any symptoms during adult life, the functions having been assumed by other portions of the brain.

The disturbance of equilibration caused by experimental destruction of the vermis of a hemisphere is manifest by a tendency of the animal to rotate on its own axis, the so-called circus movements. When the right half of the cerebellum is removed the rotation is toward the right, and vice versa. This phenomenon is not always present, but unsteadiness of gait and profound incoördination of all voluntary movement invariably result. No true paralysis occurs in cerebellar lesions, but despite this, in animals experimented upon, there seems to be a motor paresis of the limbs of the homolateral side.

The so-called cerebellar attitude of the head is inconstant; and the rotation of the head whether forward or away from the lesion has been found to be variable (Batten). Bonhoeffer has reported a forward inclination of the head in several cases. Displacement of the eyes always occurs from unilateral ablation, both eyes being turned away from the seat of injury, the eye on the opposite side of the lesion usually being more displaced than the one on the same side, and nystagmus is invariably present.

The *symptoms* of cerebellar disease may be divided into those due to injury or irritation of the cerebellum itself, those due to increased intracranial pressure, particularly pons and oblongata symptoms, and those due to sudden changes in the blood supply, which, perhaps, causes seizures.

The major symptoms are motor asthenia, hypotonus, astasia, titubation, asynergia, dysmetria, and ataxia (the latter having definite features which entitle it to the designation *cerebellar ataxia*), tremor, nystagmus, and disordered phonation and articulation. These symptoms

constitute a pathognomonic group of symptoms called the "cerebellar syndrome." The most important of these symptoms is the disorder of station and gait. The reeling, reckless gait is so familiar that description is unnecessary. The titubation or apparent constant balancing while the patient is standing is nearly as familiar. Cerebellar asynergia is the most important symptom, as it is the basis of most of the others. Babinski has shown that volitional equilibrium must be considered in two aspects: first, static equilibrium, and equilibrium of the body in a state of movement and that a lesion of the cerebellum may apparently produce the paradoxical result of very materially diminishing kinetic equilibrium, while at the same time increasing static equilibrium (cerebellar catalepsy). A patient so affected lying upon his back can hold up one or both lower extremities for several minutes without trace of tremor or swaying, which appear within a minute in a normal person who essays to do it. Babinski has further demonstrated that persons with cerebellar disease may be unable to make successive movements rapidly, such as pronation and supination of the wrist. The rapid succession of such movements, or of any movements for that matter, involves the faculty of arresting suddenly a motor impulse, *i. e.*, a contraction in a muscle or group of muscles, as well as sending to other muscles a new impulse which will cause them to contract. This faculty is essential to kinesthetic equilibrium, and for some reason not yet explicable it is disordered in cerebellar disease. The best way to display this capacity is to have the patient lay his hand flat upon the table and then rapidly to supinate and pronate the hand. Patients with cerebellar disease do it very slowly or not at all (dys or adiadokokinesis).

Tetanus-like seizures occurring in cerebellar disease, especially of the middle lobe, were first described by Jackson. They consist of exaggeration of the "cerebellar attitude" plus tonic spasm in certain groups of the bilateral muscles of the trunk and the extremities which comes on suddenly, lasts a variable time, and is not usually attended with loss of consciousness. But motor disorder, aside from asynergia in its various manifestations and dysmetria, is not an important sign of disease of the cerebellum. Cerebellar tremor has nothing characteristic to distinguish it from other intentional tremors. Paralysis and contracture when they occur are the result of ventricular distension or neighborhood lesions. The tendon-jerks may be altered in cerebellar disease, but the alteration is not characteristic or constant. Cranial nerve paralyses, of commoner occurrence with lesion of the *superior* cerebellar peduncles than of any other part of the cerebellum, and which are caused indirectly, that is by extension of the lesion, by pressure, and by action at a distance, are usually on the side of the lesion, the facial and auditory nerves being oftenest affected. The sixth, ninth, tenth, eleventh, and twelfth nerves may indicate indirect affection through impaired function. Disorder of sensibility and of the intelligence are not symptoms of uncomplicated cerebellar disease, although they are not infrequent, especially such symptoms as visual hallucinations and motor excitement.

*Cerebellar Localization.*—A certain amount of differentiation of the seat of the lesion is justifiable, especially of disease of the middle lobe and the

vermis from lesion of the lateral lobe. In lesion of the middle lobe and vermis the symptoms are equally distributed on both sides of the body; head retraction and opisthotonos are especially likely to occur. Such lesion is more frequently accompanied by paralysis of the seventh and eighth nerves, the result of indirect pressure. Sudden death in cerebellar disease from pressure upon the oblongata is more likely to occur with disease of the vermis. In lesion of the lateral lobes the muscular asthenia, cerebellar ataxia, hypotonia, asynergia, increase of static equilibrium, are present on both sides, but the symptoms are most pronounced on the side of the lesion. The head is inclined to the side of the lesion, and the face looks up away from the lesion. The patient inclines to the same side in walking and tends to fall to the same side. If rotation is a symptom, he rotates from the supine to the prone position over the side of the lesion.

**The Syndrome of Cerebellar Deficit.**—Many writers have described an association of symptoms thought to be characteristic of cerebellar agenesis, atrophy, or sclerosis. The chief symptom is disorder of motor coördination, manifest in station, gait, and speech. The usual symptoms are ataxia of "cerebellar" characteristics, with relative integrity of individual movements of the limbs when the patient is supine. He stands with the feet widely separated and legs and trunk betraying oscillations, titubations, which are sometimes so intense that he falls. Ataxia of the upper extremities and general muscular asthenia often exist in these cases. Speech is often scanning or otherwise seriously disordered and nystagmus usually exists. There may be no symptoms whatsoever. Such instances are supposed to be examples of sensorimotor cortex compensation.

Recently an attempt has been made to classify primary diseases of the cerebellum according to their pathological nature. Mangazzini distinguishes the following varieties: (1) Agenesis and atrophy of the cerebellum; (*a*) unilateral, and (*b*) bilateral; (2) Atrophy of the cerebellum; (*a*) in conjunction with disease of the brain, and (*b*) in conjunction with disease of the spinal cord. Holmes suggests the following: (1) Primary parenchymatous degeneration of the cerebellum; (2) olivopontocerebellar atrophy; (3) progressive cerebellar disease due to vascular or interstitial lesions; (4) acute cerebellar lesions, apart from tumor and focal lesions of vascular origin; (5) degeneration of the spinocerebellar tracts, the cerebellar being normal or small only; (6) cerebellar symptoms associated with a small central nervous system. Under the first are included familial and sporadic cases, occurring at any age and usually slowly progressive, with symptoms constituting the cerebellar syndrome. The morbid anatomy is a primary and progressive degeneration of the nervous elements of the cortex of the cerebellum, especially of the Purkinje cells, and of the fibres springing from the cortex.

*Olivopontocerebellar atrophy*, first described by Thomas, is characterized clinically by the cerebellar syndrome. It is neither hereditary, familial, nor congenital. It develops in late adult life and its course is progressive. It is characterized anatomically by atrophy of the cerebellar cortex and the bulbar olives, and of the gray matter of the pons; by total degeneration of the middle cerebellar peduncles, by partial degeneration

of the restiform bodies, and by relative integrity of the central nuclei of the cerebellum.

Under the third caption are included those cases of cerebellar disease incident to arteriosclerosis and primary interstitial proliferation. There is no doubt that the cerebellar syndrome may be caused by sclerosis of the bloodvessels of the cerebellum, focal or general, and the consecutive changes in the parenchyma. There are cases of disseminated sclerosis in which the sclerotic tissue is limited to the cerebellum, that may be included under this heading.

Under caption number four are included particularly acute cerebellar inflammation, hemorrhagic or otherwise, and hemorrhages into the cerebellum. It comprises also those cases in which the cerebellar syndrome comes on after acute infectious diseases, especially in children with or without symptoms indicative of encephalitis. It is important to recognize this occurrence. The fact that it may end in recovery must also be kept in mind. The relationship of this form of cerebellar disease to alcoholism, primary anemia, and toxic states has yet to be established.

Under the fifth heading are some of the cases (such as those of Sanger Brown), which Marie utilized to support the existence of hereditary cerebellar ataxia, and, whatever justification there is for adhering to the designation as a clinical convenience, it is given by these cases, in the majority of which there was no disease of the cerebellum. And as the hereditary feature is lacking in some cases, the sooner the designation is given up the better the outlook for interpreting disease of the cerebellum.

**The Symptom Complex of Occlusion of the Posterior Inferior Cerebellar Artery.**—Occlusion of this vessel causes symptoms so sharply defined that the diagnosis can be made with much certainty. The symptoms are manifold, usually coming on abruptly without disorder of consciousness. They consist of slight motor weakness of the limbs, analgesia and thermo-anesthesia in the limbs opposite the lesion, and in the distribution of the fifth nerve on the side of the lesion, usually associated with pain or paresthesia in the area in which objective sensory disturbances exist. Tactile and postural sensibility are usually attacked. Ataxia in the limbs on the side of the lesion, which bespeaks involvement of the restiform body or the cerebello-olivary fibres, is of very constant occurrence. With this there is often an inclination for the head to drop to the side of the lesion, and in some instances the patient tends to fall toward the same side. In addition to this there are symptoms indicating disturbances of Deiters' nucleus, the vestibular, cochlear, and vagus nerves, such as Ménière's syndrome, vertigo, vomiting, and auditory hallucinations. Paralysis of the muscles of deglutition and of the soft palate on the side of the lesion, is of frequent occurrence in these cases. The larynx is sometimes paralyzed on the side of the lesion, which causes disturbance of phonation. The sixth and seventh cranial nerves are rarely affected on the side of the lesion. Symptoms indicating disturbances of the sympathetic nerve supply are smallness of the pupil, narrowing of the palpebral fissure, retraction of the eyeball, and anhidrosis, all on the side of the lesion. Occasionally obstinate hiccough and

tachycardia occur. Hemiasynergy and loss of the tendon reflexes on the side of the lesion may exist.

**Lesion in the Pontomedullocerebellar Space.**—Lesion of the cerebellopontine angle, or of the space above designated, usually a tumor springing from the eighth nerve, occasionally from the fifth, seventh, and other bulbar nerves, and sometimes the accompaniment of a central neurofibromatosis, produces more or less characteristic symptoms referable to a single cranial nerve, especially the eighth or fifth, which usually long antedate symptoms characteristic of disease of the brain stem or cerebellum. Such symptoms are tinnitus aurium, Ménière's syndrome, auditory paresthesia, occipital headache, pain in the domain of the fifth nerve, and rarely facial paralysis. These symptoms are in many instances indefinite and not infrequently exist for years without incapacitating the patient. Encroachment upon neighboring organs is manifest by cerebellar ataxia, nystagmus, and occasionally paralysis of associated ocular movements, dysarthria and dysphagia. Hemiplegia may occur, usually on the side of the tumor, but it has been observed on the opposite side. The headache is by no means always occipital; frontal and crossed frontal headache have been noted. Circulatory, respiratory, and vasomotor disturbances have likewise occurred.

**Lesion of the Pons.**—Disease of the pons, both acute and chronic, is of frequent occurrence compared with disease of other parts of the brain. Inflammatory processes, acute and chronic softenings, and new-growths occur there and produce symptoms interpretation of which leads to a topical diagnosis. The symptoms vary with the site of the lesion, its size, its intensity, and the rapidity with which it develops. A slowly growing lesion, such as tumor, often exists for a long time.

The dorsal part of the pons, which contains the chief nuclei of origin of the sixth, seventh, and eighth nerves, the posterior longitudinal bundle, and not far from this ventrally, the superior olivary nucleus and the motor nucleus of the fifth nerve, and toward the mid-line the fillet, is the commonest site of disease in this region.

Disease of the pons is characterized by fairly definite symptoms, and in the books the "*pontile syndrome*" receives full consideration. It is true that lesion of certain parts of the pons produces a more or less typical symptom complex of crossed paralysis, *i. e.*, paralysis of one side of the face, which may include the tongue, some of the eye muscles, the abducens and motor oculi, and of the other side of the body, but disease of the pons covering a considerable area occurs without any recognizable symptoms.

The pons is the medium of transmission of many pathways, and the apparent origin of several cranial nerves and the termination of many nerve fibres (nuclei pontis) whose functions have not been definitely determined. Although these constituents are contained within a relatively small area, foci of the disease may be microscopic and produce extremely limited symptoms or no symptoms at all. On the other hand, a lesion that affects one-half of the pons, extending from the dorsal to the ventral border to include the pyramidal tract, may produce symptoms indicative of implication of the third, fifth, sixth, seventh, and eighth

nerves, disturbance or paralysis of associated lateral movements of the eyeballs indicating lesion of the posterior longitudinal bundle, disturbances of sensation, which may reach complete hemianesthesia due to destruction of the fillet and the ascending root of the fifth nerve, and motor paralysis due to destruction of the pyramidal tract. In addition to such symptoms, lesions of the pons adjacent to the oblongata are accompanied by profound vasomotor symptoms, flushing of the skin and hyperidrosis, which indicate that this part of the pons has an important representation for the sympathetic nervous system. Experimental destruction of half the pons in lower animals is accompanied by convulsions and coma, and similar lesions from disease in man produce similar symptoms.

When we consider that in addition to such symptoms as those above enumerated, disease of the pons of slow growth, such as tumor, causes increased intracranial pressure resulting in headache, vertigo, nausea and vomiting, choked disks, and mental disturbances, and that acute lesions such as encephalitis and acute softenings are accompanied by symptoms indicative of disturbances in other parts of the brain in addition to that which manifests the focus of the disease, some idea will be gained of the difficulty of making satisfactory localizing diagnosis in many cases. The occurrence of alternating hemiplegia will naturally establish the diagnosis of pontile disease, but these localizable lesions represent only a small number of the cases of pons affection.

An alternating hemiplegia is hemiplegia of one side of the face and the opposite side of the body. It is of two types, inferior and superior. When there is paralysis of the abducens or facial nerve on one side, and of the extremities on the opposite side, it is called the *inferior* type, whereas if the hemiplegia is associated with an oculomotor paralysis on the opposite side, it is spoken of as the *superior* type. Alternate hemiplegia was first described by Millard and by Gubler, and is frequently known as the Millard-Gubler type of paralysis. Alternating paralysis of the inferior type is sometimes complicated with involvement of the tongue, and dysarthria is a conspicuous symptom. There is difficulty in articulation, and in some instances difficulty in swallowing. These symptoms constitute *pseudo-bulbar paralysis*. If disturbance in the associated movements of the eyes on one side is added to such a clinical picture, the topical diagnosis is fairly certain, because such symptoms indicate that the lesion involves the posterior longitudinal bundle. In such cases the lesion must be predominantly of the dorsal part of the pons.

Absence of hemiplegia, be it alternating or otherwise, does not exclude lesion of the pons. Partial or complete hemianesthesia, hemiataxia, cerebellar ataxia without hemiplegia, may occur with lesion of the pons, and may be deemed to be of such a nature when they are associated with paralysis of one of the motor cranial nerves having their origin in the pons or the ascending root of the fifth nerve, and which cause alternating hemiplegia, insensitiveness of the cornea, and half-sided disturbances in the associated movements of the eyes. Lesion of the pons may cause motor hemiplegia with no other symptoms when the lesion is in the ventral part of the pons, of small size, and confined narrowly to the pyramids. It is said that such pontile hemiplegia causes more profound



involvement of the trunk muscles than capsular hemiplegia, and that the face is usually spared.

A lesion developing ventrally in the pons may, by pressure upon the posterior part of the oculomotor nuclei, cause paralysis of associated upward movement as a result of injury to the nuclei over the superior rectus and inferior oblique muscles. Paralysis of downward associated movements depends on impairment of the inferior rectus and the superior oblique muscle, which have cells of origin in two distinct nuclei. A lesion of the nuclei of the inferior rectus muscles and of the fibres connecting them with the nuclei of the trochlear nerves causes paralysis of associated downward movements.

Conjugate deviation of the eyeballs from pontile lesion is uncommon when the lesion is confined to one side of the pons. It has been stated repeatedly that conjugate deviations of the eyeballs away from the site of the lesion is common in unilateral pontile lesion. It is probable that in most cases the side of the pons opposite to the lesion is directly or indirectly involved in these cases. Pons lesions almost always cause pupillary symptoms and tonic convulsions in all extremities, also in the face, with the exception of those areas which are innervated by nerves coming from the pons whose angle or whose intrapontile course has been destroyed or disturbed by the lesion.

*Superior Pontile Syndrome.*—Raymond and Cestan describe a superior pontile syndrome characterized by paralysis of the lateral associated movements of the eyes and paralysis of the arm and leg with the characteristics of a central paralysis, the inferior facial being affected on the same side. The motor symptoms were manifest particularly by trembling, athetosis, incoördination, and asynergia. All forms of sensibility, stereognostic, muscular, articular, and postural, were affected. The eyes were fixed straight ahead, and the pupils reacted, *i. e.*, there was paralysis of the voluntary movement of the sixth pair and paralysis of the associated lateral movements of the eyes. This was associated with preservation of the power to converge and integrity of the movements of elevating and lowering the eyes, known in France as the “*Syndrome of Parinaud*,” a quasi-pathognomonic sign of lesion of the superior part of the pons. The lesion was a tubercle of the right half of the pons affecting the median fillet, destroying the fibres going from the cortex to the sixth pair of the fibres which are supposed to unite the nuclei of the third and the sixth pairs.

**Pituitary and Pineal Glands.**—Disturbance of the function of the pituitary gland is evidenced by a syndrome of which acromegaly, adiposity, stunted growth, and maldevelopment of the genital organs are the chief manifestations. The former is believed to be the result of hypersecretion, the latter of hyposcretion of the epithelial part of the gland. The commonest cause of disturbed function of the gland is tumor. Excessive growth of hair and deposition of fat, associated with abnormality of the genitalia, suggest disease of the pineal gland. Such symptoms followed by bilateral ocular paralysis, nystagmus, abnormality of the pupils, and ataxia constitute the *pineal gland syndrome*.

## CHAPTER VIII.

### APHASIA.

BY JOSEPH COLLINS, M.D.

THE most interesting association of symptoms in the topical diagnosis of disease of the brain is that known as aphasia. The externalization of thought in any form, and its communication to another is a psychophysical act of great complexity. When this capacity is disordered or destroyed in an individual, the phenomena that accompany such disorder or destruction are very striking. In attempts to determine the localization of the speech faculty we have had to rely entirely upon the mode of investigation known as the pathological method, that is the study of symptoms first and the seat of disease later. The result has been that more has been taken for granted on inadequate evidence or insufficient proof in the localization of the speech faculty than in the localization of any other distinctive function.

*Aphasia* is a term used to indicate any disturbance or perversion of intellectual expression. It includes all defects or disorders of intellectual expression, whether the result of disarrangement or destruction of the receptive or of the emissive components of the speech mechanism, or of anything which may be employed as the substitute or equivalent of speech. It may be the result of conditions by which the patient is unable to part with the expressive equivalent of an idea which has been properly formed; it may be caused by any condition that interferes with the reception of impulses or stimuli that enter into the genesis of ideas to be used in the construction of internal or external language. As movement in some form is requisite for the manifestation of all expressions, defect of this is the condition to which the term *motor aphasia* or aphasia of emission has been and is still often applied. In the second form of aphasia the sufferer is unable to adapt receptive communications and make them fit the idea represented by the verbal symbol, auditory or visual; that is, he has lost the faculty of adapting the complement of the word to his own idea; it matters not whether these words be spoken or written, or communicated by some equivalent, such as music and pantomime. In a general way, this is aphasia of reception or *sensory aphasia*.

Aphasia is a symptom; not a disease. It may be classified as: 1. *True Aphasia*.—Aphasia of apperception. Due to lesion of any constituent of the cortical speech area, constituting the zone of language.

2. *Sensory Aphasia*.—Due to lesion of the central and peripheral sensory pathways leading to the zone of language.

3. *Motor Aphasia*.—Due to lesion of the motor pathways, over which the motor impulses travel, in passing to the peripheral speech musculature.

4. *Compound Aphasia*.—Any combination of two or more of these.

If this classification were adopted, it would simplify the matter enormously, inasmuch as there would be in reality only one aphasia; moreover, it would harmonize with the clinical manifestations. It will be seen that the scheme is not unlike that which has been set forth by Marie. He denies No. 2 and calls No. 3 anarthria or aphemia. Some points in the history of aphasia facilitate an understanding of this difficult subject. There are three important epochs.

1. The publication in 1861, by Broca, of an example of aphasia and a description of the brain of the individual, which seemed to prove that the lesion was of the posterior part of the third frontal convolution.

2. The classical work of Wernicke, in 1874, which demonstrated that lesion of the first temporal convolution caused a definite symptom complex, from that time described as "sensory aphasia." Bastian had anticipated Wernicke in many of his contentions and conclusions.

3. The papers of Pierre Marie, entitled "Revision of the Question of Aphasia" in 1906, and the studies of Liepmann, of Berlin, on apraxia.

**The Zone of Language.**—This is the name given to the area of the brain in which are carried on the processes essential to speech and its components. This zone is not to be defined narrowly. It varies in individuals and at different periods of life in the same individual, *i. e.*, it is susceptible to phylogenetic, and to ontogenetic variation, the latter depending somewhat on the speech acquisition of the individual, and on the range and number of avenues by which he receives or has schooled himself to receive information.

The speech area, or zone of language, is an area made up of neurons, some of which send their axons into the Rolandic region and into the frontal regions of the brain, while others confine their distribution to the speech area itself; and, as they do not pass outside of this area, they may be looked upon as intercentral neurons.

The centres of speech in the area of language are three in number: a centre for auditory memories, for visual memories, and for articulatory-kinesthetic memories. The latter is often called the centre for motor memories. The validity of this teaching is under discussion. These three centres have definite localization. The centre in which are stored the memories of articulation is situated in the third frontal convolution, immediately adjacent to that portion of the Rolandic cortex the cells of which give origin to the projection fibres going to the tongue, the lips, and the larynx. The centre in which are stored the visual images is situated in the posterior portion of the inferior parietal lobule. The calcarine cortex is the percipient cortical visual centre. It embraces part of the dorsal lip, the bottom and the ventral lip of the calcarine fissure from the region of the peduncle of the cuneus to the posterior lip of the retrocalcarine fissure. It extends on the lingual gyrus and on the lower part of the first occipital gyrus. The geniculocalcarine path is its afferent radiation. If we have in mind the afferent radiation or central projections of the optic tract after they leave the external geniculate body, the inferior quadrigeminal body, and the pulvinar of the thalamus, until they reach the lingual and fusiform lobules bordering the calcarine fissure,

we shall appreciate that the angular gyrus is the most direct, the most adjacent, and the most elective place in which the visual images could be stored. In fact, its relationship to the primary visual centre and to the fibres that convey visual impulses, the radiations of Gratiolet, is analogous to the environmental relationship of the centre for articulatory memories and the Rolandic cortex that externalizes speech.

The third centre, the auditory centre, the most important of all the speech centres, is situated between these two centres in the zone of language, and occupies the first temporal convolution, and particularly that portion of the cortex which surrounds the temporoparietal sulcus. The centre for the storage of auditory memories is in a definite part of the general auditory area, in the posterior part of the first temporal convolution immediately adjacent to the gyrus in which are stored visual memories. Thus, it will be seen that the auditory and visual memories, which are contributory to the development of speech and education in general, are immediately adjacent, constituting one continuous area.

Marie maintains that there is no trustworthy clinical observation that substantiates the claim that the first temporal convolution is the centre of hearing, but his views on this phase of the matter cannot be accepted. Probably there is no fixed spot in the first temporal convolution that is the sole repository of auditory memories. The cortical auditory area may vary in different persons, but anatomical researches by Flechsig, by Brodmann, Meyer, and others, demonstrate conclusively that the transverse temporal gyri and the adjoining part of the first temporal convolution are decidedly richer in acoustic fibres and have a distinct connection with the internal geniculate body. The economy of Nature is such that anatomical possession entails functional endowment.

It is folly to speak dogmatically at the present time of the relationship of sensory aphasia and its various clinical components to lesion of the auditory receiving station. Until there is a record of many cases carefully observed clinically, and until we are fortunate enough to encounter cases in which the symptoms carefully noted are found to depend upon small and single lesions, such discussion as that carried on by Moutier, defending the Marie theory, and Ladame and v. Monakow, leads nowhere. The relationship of the auditory word-perceiving centre to the word-elaborating area is probably a close one, but just what it is cannot be said dogmatically. Destruction of the left anterior transverse convolution produces in right-handed persons word deafness. Whether it might be affected to such a slight extent as to cause partial word deafness and such slight affection of internal and articulate language as to be detected only with difficulty seems quite probable.

**Clinical Classification.**—It has been customary to divide aphasia into (1) motor aphasia, (2) sensory aphasia, (3) total aphasia. Each of these is further subdivided, the second into auditory and visual aphasia. The first two varieties have always been considered distinct, although it has been generally admitted that it is not always easy to distinguish one variety from another. Anatomically speaking, sensory aphasia might be defined as aphasia due to a lesion of the posterior part of the area of language, and motor aphasia as due to the anterior end of this zone.

The subcortical forms occur when there is lesion of the pathways which carry impressions into and away from the zone of language.

**So-called Motor Aphasia.**—This has been classified as (*a*) cortical, due to lesion of the foot of the third frontal convolution, which abolishes the “memory images” of articulation, and (*b*) subcortical motor aphasia (pure motor aphasia of Dejerine), due to lesions of the motor pathway over which speech impulses or messages travel to be externalized. Speech is properly formed, but it cannot be produced because of the hiatus that exists in the speech pathway. These two varieties may often be readily distinguished. But it must be said that the features, which allow us to distinguish the one from the other, are not quite so absolute and convincing as one might be led to infer from reading works on the subject. Proust and Lichtheim suggested a test for subcortical motor aphasia to prove that patients preserve the memorial motion of the word; that is, that they have in their minds the name of the object, which they are incapable of emitting. To test this deficiency of internal language, the patient who hears a polysyllabic word, or sees and recognizes an object indicated by that word, is asked to indicate by some movement the number of syllables; then to indicate by similar pressings the number of letters in the word and the number of letters in the syllables.

Cortical motor aphasia, Broca's aphasia, is characterized by a loss of spontaneous and repeated speech, some disturbance of the capacity to read articulately, slight intellectual enfeeblement and by preservation of the capacity to comprehend articulate speech. In some instances interpretation of spoken speech is slightly impaired in true cortical motor aphasia. The peripheral speech mechanism—the tongue, lips, palate, and vocal cords—is in condition to functionate. The only justification for the use of the word motor in this form of aphasia is that the images of articulation are called into being by movement and are externalized by movement. In true cortical motor aphasia there exists the same inability to call into existence the sensory memories of articulation, and thus to make them a part of internal speech, as there is to externalize them in the shape of articulate words. Many of the cases of aphasia in the literature, which are considered to belong in this category, are not of this variety at all, but are examples of subcortical aphasia; that is, disturbance of speech dependent upon interruption of the projection tracts, which convey the articulatory impulses from the cortical area of the peripheral speech mechanism to the peripheral speech apparatus.

Associated with this loss of spontaneous speech there is an inability to repeat words and to read aloud, but the patient comprehends spoken words, oftentimes imperfectly. There is inability to express thoughts in writing, because in writing the motor word representations are always revived by the impulse which travels from the percipient centre (which is either in the visual area of the brain in spontaneous writing, or the auditory speech area in writing from dictation) through the articulatory kinesthetic centre to that part of the Rolandic region which guides the part of the body holding the pen. Incapacity to write is proportionate to the amount of derangement of internal language, and it bears a definite relation to the amount of latent or actual visual amnesia of words which

every patient with cortical motor aphasia has. In most cases the capacity to write is limited to the name and a few other words that have been done so habitually, automatically, and frequently, that they form a part of patient's habitual acts, and are done almost reflexly. Writing voluntarily and writing from dictation are practically impossible, yet the patient is able to write from copy.

The patient is unable to call up promptly and vigorously auditory images; that is, he has some word deafness, and in many instances amimia. This word deafness becomes conspicuous when the patient is spoken to abruptly and rapidly. The capacity for articulate expression which some motor aphasics retain is for a few words whose utterance partakes more of the nature of a reflex act or of an emotional possession than it does of a process of intellection. Although in Broca's aphasia the power to make voluntary expressions is usually entirely gone, the loss may be partial. When partial, the power of expression is limited as a rule, to monosyllabic words, usually verbs. Occasionally patients who are afflicted with complete motor aphasia are able to utter some words of the nature of an oath, which seem to escape from them in a rapid, uncontrollable way, or to ejaculate words expressive of the feelings. Such expressions are not the product of cognition, but of the emotions, and partake of the nature of reflex action. Other patients repeat continually some expression or meaningless word or words. Such recurring utterances are distinctive features of cortical motor aphasia and not of the subcortical variety.

Cortical motor aphasia, due to small lesions, is sometimes manifest merely by a loss of substantives, amnesia of the names of the things or objects of which the patient tries to speak, by inability to construct sentences correctly, and by paraphasia.

The symptoms of subcortical motor aphasia are practically the same as those of Broca's aphasia, with two important exceptions; the patient retains the capacity to write and to read, and he responds to the Proust-Lichtheim test. In other words, the patient with subcortical motor aphasia retains the capacity to talk to himself, to speak without words. He hears, sees, writes, mimics, and in other ways gives evidence of intellectual integrity and internal language. He is incapable only of articulate speech. This may not be entirely lost. Indeed, it varies through dysarthria, dysrhythmia, up to complete anarthria and arrhythmia, and thus to complete speechlessness. In the conventional usage of the term this is aphasia. But it is not true aphasia, for true aphasia occurs only with lesion of the area of language, and is invariably attended with disturbance of internal language.

**Sensory Aphasia.**—This is the designation given to the speech disturbances or imperfections of language due to lesion of the posterior part of the speech area, more particularly the temporal and angular gyri; that is, to lesion of the perceptive areas of the brain and the immediate incoming special sense and commissural pathways of such areas. The perceptive centres by whose functioning speech is ontogenetically developed are the auditory and the visual, and sensory aphasia is thus practically auditory and visual aphasia. It may be defined as loss of the under-

standing of words due to interference with the formation of associations necessary for complete perception. Sensory aphasia differs materially from motor aphasia. In the first place it is not usually accompanied by hemiplegia unless the underlying lesion is very extensive. It is the aphasia of comparative speechfulness in contrast with the speechlessness of motor aphasia. It is characterized by logorrhœa, motor aphasia by alogia. The course of sensory aphasia is rather typical. The patient starts with senseless loquacity and gradually his vocabulary shrinks, often reaching frank mutism when the auditory area is completely destroyed.

When the lesion of the auditory centre is slight, the patient may *seem* to understand what is said to him but he cannot use words with their proper signification and paraphasia may be the distinctive symptom. Jargon-aphasia, or the production of a jumble of words all forged into one, the syllables of which may be articulated, but the words have no similarity to words as usually spoken, may be looked upon as an extension of paraphasia.

The degree to which spontaneous writing may be preserved or lost in sensory aphasia varies with the patient and with the seat and intensity of the lesion. Incapacity to write is most striking when word blindness is prominent and total agraphia always accompanies destruction of the angular gyrus. If the lesion is predominantly of the auditory area, there is inability to write from dictation. Spontaneous writing may be preserved to a considerable extent, but the output is usually senseless and disordered. The capacity to copy is preserved, but the patient makes an exact reproduction of what is before him. This is especially true of cases in which there is lesion of the angular gyrus.

In sensory aphasia with word deafness there is inability to understand spoken words. This is dependent apparently upon the total loss of auditory verbal memories. It is one of the commonest forms of aphasia, and it rarely occurs independently, being frequently associated with some degree of word blindness or manifestations of Broca's aphasia. In sensory aphasia with word deafness the patient hears the voice with which the words are spoken, but they contain no meaning to him. He often recognizes the significance of other sounds, unless it be that the memory pictures of such sounds are also lost. Naturally these different degrees of word deafness depend upon the extent of the lesion or the destruction of the auditory area. The appearance of such patients is very significant. They are quiet and observant; their glance betrays unawareness or suspicion, and their demeanor is often one of restlessness. This alteration of manner, the inability to repeat what is said to them, and the profound diminution of spontaneous speech often cause them to be looked upon as demented.

To the form of aphasia in which there is deafness for musical notes the designation tone deafness is given. Musical deafness is almost always associated with word deafness, but there have been a few cases recorded in which it occurred apart from the latter. Many attempts have been made to classify the clinical varieties of amusia, but to no purpose.

One of the commonest manifestations of sensory aphasia is loss of the

verbal memory of written and printed symbols—*word blindness*. It is of constant occurrence when the lesion is of the posterior part of the area of language. The visual area is made up of a perceptive centre situated on the mesal surface of the occipital lobe along the calcarine fissure, and of the centre in which is “stored” the visual memories of words, other graphic symbols and probably of objects, situated in the posterior portion of the inferior parietal lobule, the angular gyrus, and the adjacent margin of the supramarginal convolution, which curves over the posterior extremity of the fissure of Sylvius. It must be admitted that the localization of the “visual word centre” is based largely on theory and slightly on facts. Destruction of this centre causes word blindness or alexia and agraphia, but no loss of visual acuteness. The primary visual area and the higher visual centre are frequently diseased simultaneously, and when this occurs homonymous hemianopsia due to lesion of the primary visual area is superadded to the word blindness. There are various degrees of intensity of word blindness in sensory aphasia. The patient may be unable to read words and yet retain the faculty of recognizing letters, or he may be able to recognize letters and be unable to join them in syllables—asyllabia it is called. The patient may not be blind to all forms of notation, graphic and symbolic representation. Thus there may be sensory amusia, sensory asymbolia, sensory amimia, etc. In many cases with word blindness there is preserved the capacity to recognize certain familiar words, such as the patient’s name or words that he has been accustomed to see frequently. There are also cases in which numbers are recognized, but letters are not.

If the angular gyrus is completely destroyed the patient has *agraphia*. In those cases in which voluntary writing is preserved, the lesion involves the primary visual centre, and the patient has right homonymous hemianopsia; the patient begins to write at the extreme left side of the sheet and stops in the middle of the page. The patients, being unable to read what they have written, are unconscious of any errors in spelling or phraseology that they may make, although they put the words on paper in an orderly fashion. Patients with word blindness are sometimes able to read written or printed words by tracing the word with the index finger or a pencil, thus substituting kinesthetic for visual memories, which kinesthetic memories revive the mental concept of the word.

There has been a very great amount of analysis of *visual aphasia*, as that form of sensory aphasia in which word blindness predominates is often called, and much speculation has been indulged in as to the pathological physiology and psychology of these subdivisions. There is a variety to which the name verbal *amnesia* or psychic blindness is given. The patient interprets letters as letters and words as words, and he can read them and copy them, but they convey no meaning to him. When they are pronounced by someone else he hears and interprets them readily, but he has no idea that they are the same words that he has been reading or copying. The lesion which it is supposed produces such a condition is one that interferes with the pathway that conveys the sensation from where the memory of the printed word or object is stored to the place where the idea is formed, if there is any such special place.



Another variety is that to which the name *optic aphasia* has been given by Freund. The patient, on looking at an object which he has previously seen or used, is unable to call up its name, although he is able to utter it when it is recalled for him. The lesion is supposed to be one that interrupts the pathways that unite the seat of cortical visual representation and the seat of cortical auditory memories.

A third variety is that to which the name of *psychic blindness* or mind blindness is given. It is the *Seelenblindheit* of the Germans and the *cécité psychique* of the French. In this condition the patient not only does not recognize the significance of letters, but he loses the power to differentiate between objects or persons and to distinguish the use of things; in other words, it is word blindness plus apraxia. The word *apraxia* is coming to have, or it may be said has come to have, a wider application than was formerly given to it. The term was used to indicate inability to comprehend the usage of ordinary objects and things to which one has been accustomed. It was thought to be due to the abolition of the visual memories of objects which are stored in the cortex of the parietal lobe adjacent to the centre for verbal memories. The word *apraxia* is now used to indicate inability to perform certain familiar purposeful movements by one who has neither paralysis nor ataxia. *Apraxia* may occur without aphasia. According to the accepted doctrines of aphasia of today, aphasia may occur without *apraxia*, but if Marie's ideas of aphasia are accepted, so-called motor aphasia will be looked upon as a form of *apraxia*, that is, *apraxia* of the speech musculature.

It is customary to describe a subcortical form of sensory aphasia analogous to the subcortical form of motor aphasia, although its delineation is not so distinct. If we divide sensory aphasia into auditory and visual aphasia, we have to divide subcortical and sensory aphasia in the same way, for anything that causes an interruption of the visual and auditory pathways subcortically will give rise to these varieties of aphasia. Subcortical word deafness is characterized by inability to understand spoken words and by inability to write from dictation. It is distinguished from cortical auditory aphasia by the preservation of spontaneous speech and by the ability of the patient to read aloud, to copy, and to write. As in every other form of subcortical aphasia internal language is intact.

The symptoms of subcortical visual aphasia vary somewhat with the seat of the lesion, that is, with its proximity to the angular gyrus. As a rule, there is word blindness, usually partial, associated with the right-lateral homonymous hemianopsia. Spontaneous speech is usually well preserved. The patient is able to write voluntarily and from dictation, but he cannot read what is written either by himself or by others. There is very slight or no word deafness.

SUMMARY.—In brief, then, we have two distinct aphasias—motor aphasia (Broca) and sensory aphasia (Wernicke); two subdivisions of each of them, and total aphasia, when the two chief varieties occur simultaneously. A patient with Broca's aphasia cannot speak, or is reduced to a word or two, he writes as badly as he speaks, and he often has some word deafness. He cannot talk to himself, and he cannot read to himself. The patient with sensory aphasia, on the other hand, is a babbler. He

uses words senselessly, jargon-aphasia, or in a preverse way, paraphasia. There is marked difficulty of comprehension of spoken or written language and profound disorder of reading and writing. In Broca's aphasia the trouble is predominantly of articulation and of writing, that is to say, in the omission of words; in sensory aphasia all the elements of language are disordered.

Broca's aphasia is therefore the suppression or profound alteration of the speech not dependent upon paralysis, associated with trouble of internal language, of speech, and of writing, dependent upon lesion of the foot of the third frontal convolution, which lesion causes destruction of "motor verbal images," or upon interruption immediately beneath the cortex of the fibres or pathways that convey motor speech impulses. Sensory aphasia is due to the lesion of the posterior two-thirds of the zone of language, and especially the first temporal convolution and the angular gyrus.

**Revision of the Aphasia Question Proposed by Marie.**—This, in brief, was the status of the aphasia question when Pierre Marie assailed it in 1906. Marie maintains that the third frontal convolution of the left side does not play any special rôle in the function of language; that which is called motor aphasia or Broca's aphasia is anarthria plus aphasia; that the aphasia of Broca is not a disease, not a clinical entity, but a syndrome, a superimposition of aphasia upon anarthria, or, better, a simple juxtaposition of two distinct troubles, anarthria and aphasia. As to aphasia itself, Marie holds there is only one aphasia, which he proposes to call Wernicke's aphasia, and only one speech centre diffusely localized in the left temporoparietal lobe, and that this centre is a region of intelligence specialized for language, not a centre of sensory images. The clinical splitting up of the aphasia of Broca into two elements, anarthria and aphasia, Marie maintains is verified by autopsy. One finds constantly lesion of the lenticular zone associated with lesion of the zone of Wernicke.

Marie's description of his lenticular zone is as follows: If in a horizontal section of the brain one carries a line in a transverse direction from the anterior fissure of the island of Reil as far as a corresponding point in the lateral ventricle, and another transverse line from the posterior fissure of the insula to a corresponding point in the lateral ventricle, a region is circumscribed having nearly a quadrilateral outline and containing in its territory the caudate nucleus and the lenticular nucleus, the external capsule with its different parts, the cortex of the island of Reil, and the internal capsule. It is in this territory very distinctly separated from the third frontal convolution that the lesion which determines anarthria especially is situated. This territory the writer designates, for the sake of brevity, the lenticular zone. The superior and inferior limits of this zone it is not yet possible to define.

*Anarthria* (or *aphemia*, the term Marie is now willing to use) is characterized clinically by the loss of speech with preservation of the understanding of words, of reading and writing. It is produced by a lesion in the lenticular zone, interfering with the coördination of movements required for the phonation and articulation of words, without introducing

true muscular paralysis. Broca's aphasia is produced by the combination—the proportion varying with the case—of the lesion of anarthria with a lesion of Wernicke's zone or the fibres coming from this zone.

The third left frontal convolution plays no special part in the function of speech. The true speech centre is the zone of Wernicke; which must be considered not a *sensory* centre, but an intellectual centre. Wernicke's zone consists of the supramarginal and angular gyri and the feet of the first two temporal convolutions. Lesion of this centre determines in proportion to its extent, and in addition to the disturbances of speech, deficient understanding of spoken words, inability to read and write, as well as the disappearance of certain concepts of a didactic character. The foot of the first temporal convolution cannot be said to constitute a sensory centre for the auditory image of words. Pure deafness does not exist. Pure alexia (pure word blindness of authors) does not occur clinically. The lesion producing it is a lesion of the posterior cerebral artery, not of the sylvian artery, as in the other aphasias. It is useless and inaccurate to drag in the angular gyrus, which cannot be recognized as the centre of visual word images.

Marie maintains that there is no reason to preserve the classification of aphasia into cortical and subcortical forms. As a matter of fact, aphasia due to focal lesions is never exclusively cortical. It is, moreover, advisable at present not to refer to the cerebral cortex the entire pathological physiology of aphasia, since the subjacent white matter seems to play a part of perhaps greater clinical importance than the gray matter. If one insists upon classifying aphasia, the varieties of which are connected by a scale of innumerable transition forms, the best division would be into (1) *intrinsic* aphasia, in which Wernicke's zone or the fibres coming from it is directly and considerably affected by the lesion (Broca's aphasia, Wernicke's aphasia), and (2) *extrinsic* aphasia, in which Wernicke's zone with its fibres is not directly involved.

For Marie there is only one aphasia, the aphasia of Wernicke. The term sensory aphasia, he thinks, should disappear. The aphasia of Wernicke has for its foundation lesion of the zone known by his name. Intrinsic aphasia is accompanied by trouble of the internal language and by intellectual deficit. The other alterations of language, anarthria, and pure alexia, are extrinsic syndromes. Pure alexia is dependent upon lesion of quite another vascular territory than aphasia. Aphasia is dependent upon lesions of the sylvian artery, and alexia upon obliteration of the posterior cerebral artery. He maintains that in autopsies upon patients with Broca's aphasia there is a double lesion. One causes the anarthria, and the other the trouble of internal language, of reading, and of writing. The terms Broca's aphasia, ataxic aphasia, total aphasia, designate the progressive degrees of the same syndrome.

The three important factors in the discussion are: (1) Is Broca's area (the foot of the third frontal convolution), the centre in which are stored the memories of phonetic speech, articulatory kinesthetic memories? In other words, is motor aphasia or Broca's aphasia real aphasia at all? If there is a well-defined, though not sharply marked, syndrome to which the name of Broca's aphasia is given, upon what is it dependent

anatomically? (2) Do lesions of the so-called lenticular zone give rise to a symptom complex parallel to that of so-called subcortical motor aphasia, the anarthria or aphemia of Marie? (3) Is the area of language the area in which memories and words seen and heard are stored, and from which alone they can be evoked by peripheral stimuli, or is the storage of such memories a function of the anterior pole of the brain (the so-called psychic sphere) or of the whole brain itself?

It must be granted that there is no adequate clinical or anatomical evidence for considering Broca's centre to be the seat of memories of articulation. There does not exist in the literature a case of Broca's aphasia clinically in which the lesion was confined narrowly to the foot of the third frontal convolution, nor does the brain upon which Broca bases his original thesis, which is now preserved in the Musée Dupuytren, which the writer has examined carefully, show it. Although there are some cases recorded in which macroscopically lesions seem to be closely confined to Broca's convolution, macroscopic examination is entirely inadequate in these cases, save that it gives an accurate idea of the extent of the lesion. Serial sections alone can be relied upon.

It may be stated that our present conception of the zone of language must be modified in so far as denying the existence of the storage of articulatory or kinesthetic memories in the anterior pole, the foot of the third frontal convolution.

It is admitted by everyone that there is an aphasia, which is clinically distinctly characterized, to which the name motor aphasia or Broca's aphasia is given. Its symptomatic features have been described. Is it sensory or Wernicke's aphasia plus anarthria or aphemia? If it is true that the foot of the third frontal convolution is not the seat of phonetic memories, then Marie's explanation is probably the correct one. Marie admits that Broca's aphasia and Wernicke's aphasia are clinically two distinct varieties. The distinction between the two is not always very sharply drawn, but, as a rule, certain characters permit the distinction to be made. Those who have contended that the symptomatology of the two forms of aphasia was very unlike have pointed out that it is a mode of copying that distinguishes motor aphasia from the sensory aphasia. It is admitted by everyone that the motor aphasic has some word deafness and some word blindness, but it is maintained that he copies print in script while the sensory aphasic copies servilely. But there has been important testimony to show that this is not true (Souques).

In regard to the *mental defect* of aphasic patients, whether they have sensory or motor aphasia, this has received due consideration in every treatise on aphasia, and there can be no doubt of its existence; considering the lesion of the brain in the majority of cases of aphasia, the wonder is that the mental defect is not more pronounced.

That lesion of the so-called lenticular zone gives rise to the symptom complex parallel to that of so-called subcortical motor aphasia there can be little doubt. The preservation of internal speech is the distinctive feature in each of them. Whether the loss of speech capacity or destructive speech capacity which results is called anarthria or aphemia is not of prime importance.

In the discussion of aphasia, Marie concedes the word *aphemia* to express speech defect of Broca's aphasia, which he first proposed to call *anarthria*, making sacrifice of the word, not of the fact. The writer agrees with Marie that the term aphasia should be reserved for those cases in which internal speech is disordered. Those cases in which the internal speech is not disordered and in which there is inability to speak, pure motor aphasia (Dejerine), should be classified as cases of *anarthria* or *aphemia*. This *anarthria* may exist from the onset of the patient's illness, that is, there may never be any true aphasia associated with it.

The answer to the third question cannot be given positively at this time. It will be appreciated that the phrase "storage of memories," which is constantly used symbolically in writing on aphasia, is not to be taken literally. We do not know how memories are created, invoked, reinvoked, or localized, or indeed that they are localized at all.

The position taken by Marie is not so revolutionary as has been commonly supposed. In the first place, it has never been seriously denied that there is a certain amount of dementia in practically all cases of true aphasia. This mental defect varies in different cases, but it has been generally recognized. In the second place, the *anarthria* or *aphemia* of Marie is in reality the same thing as the subcortical motor aphasia of Wernicke and the pure motor aphasia of Dejerine. The important contribution that Marie has made is that the foot of the third frontal convolution (Broca's convolution, so-called) is not the seat of articulatory kinesthetic memories, and is not the integral part of the zone of language, and that, therefore, destruction of it does not cause aphasia. It is not enclosed in the quadrilateral area called the *lenticular zone* by Marie.

It is probable that Marie is correct in assuming that the third frontal convolution is not an area in which are stored memories of articulation. The criticism that may justly be made on Marie's revision of aphasia is that he has torn down without building up. When he attempts to define his quadrilateral or lenticular area, we see that, although he gives an anterior and a posterior border to it, it includes in reality an enormous amount of brain, especially in the superior and inferior direction, and Dejerine's criticism that it is not a localization at all is a very just one. Whether we accept the classic teaching or not (and certainly it cannot be fully accepted after all the evidence has been considered), we must admit that Marie's service has been very great, for it has put us on toward fact and away from theorizing.

**Diagnosis.**—The speech faculty consists of two parts, the receptive and the emissive; either may manifest the predominance of aphasic symptoms. In true aphasia, that is, aphasia dependent upon lesion of the zone of language, neither can be the sole medium of manifestation of the speech defects. Emissive speech is manifest by articulation, by writing, and by pantomime. Integrity of the receptive side of language is commensurate with the interpretation of visual and auditory stimuli.

The attitude, the demeanor, the conduct of the patient may be of the greatest service from the very beginning of the examination. The manner and expression of one with sensory aphasia (Wernicke's aphasia) are frequently those of a person who has lost interest in his surroundings,

and his attitude is that of a deaf person slightly demented. Moreover, patients who have this form of aphasia are often garrulous, and on the slightest provocation, or without provocation, emit a string of sounds that convey no meaning. This is especially true of recent cases. Patients with motor aphasia (Broca's aphasia) and with subcortical motor aphasia (anarthria or aphemia of Marie) are very different. The aphemic is often absolutely silent, but watchful, and the intensity with which he holds every move of persons around him is very striking. He may be absolutely speechless, yet capable of understanding all that goes on about him and within his hearing and vision.

A number of schemes have been devised to facilitate the examination of aphasic patients, and the following simple plan is found most serviceable. After securing a general history of the patient's life and of his previous illness from some member of the family, and in this way getting information of the character of the disease of which the aphasia is a symptom, the patient's ability to express ideas, to receive and interpret information through every avenue should be tested. The mental processes, apart from the manifestation of mental states, and the mental capacity for the reception of sensory stimuli, then should be examined. Although a number of these may be determined simultaneously, it is best to take each one separately, such as attention, memory, orientation, capacity for retaining impressions, for deliberation, for reasoning, and his temperament or mood.

In approaching a patient with aphasia, it is natural that endeavor should be made to elicit information by speaking to him. It becomes necessary, therefore, to determine if the patient takes note of what is said to him orally, and secondly, if he understands what is said. In other words, does spoken speech awaken in his auditory centre and in the auditory interpretative area corresponding memories? This can be done ordinarily by taking some simple question, as, "How long have you been sick?" or by addressing to him some simple command, such as, "Give me your hand." Care must be taken not to employ too conventional questions or commands, such as, "What is your name?" and "Put out the tongue." The patient may have lost the auditory apperceptive faculty and still make reasonable reply to such questions merely from association and habit. Naturally, the patient should not get any information of what is being asked through any other avenues than those of hearing. Such patients are quick to grasp, particularly if they have been aphasic for some time, the significance of even slight emotional expression or pantomime on the part of the interlocutor. If the patient does not reply to such questions or commands, there may be trouble with the receptive or with the emissive speech faculties. If he is word deaf (that is, if the trouble is one that prevents the sound of the word from reaching the centre in which the memories of previous word sounds are said to be stored), the patient will not endeavor to respond by word or act, though in some instances he does so. Nor will the face show the slightest response or indication of comprehension. If he does respond, the diagnostic feature is that his answer, even though it be made up of articulate words, has no pertinency or bearing on the question. If the patient is

not word deaf, he will make some movement, be it of the head, hand, or features, to indicate that though he understands, he cannot reply. Generally, this gesture is very significant. It consists of a despairing expression of the countenance and a touching of the lips or the throat with the fingers. Oftentimes the question can be decided very quickly, if there remains some doubt even yet, by asking some absurd or ludicrous question and noticing how the patient receives it. If, in reply to the question, "Are you one hundred years old?" he solemnly says "Yes," or if he does not see the ludicrousness of a request to turn a somersault when he is obviously paralyzed, it is rather convincing proof that such speeches do not awaken the proper responses in his mind, and if there be no dementia, it is suggestive evidence that the patient is word deaf, and the examination should then proceed from that standpoint. It must be borne in mind that there is a varying degree of word deafness in cases of motor aphasia. Thomas and Roux suggest a method for eliciting it. The patient is shown an object and the examiner pronounces several syllables of the name of the object, the first, last, or intermediate syllable. In minor degrees of word deafness the patient recognizes some of the syllables, not all.

Although all the speech centres may be simultaneously disorganized, the symptoms attributable to lesion of one usually dominate the speech defect. If the examination so far seems to suggest the existence of the word deafness as the leading feature of the sensory aphasia, it should then be determined to what degree of completeness this exists, and the extent and kind of disturbance that it causes in the externalization of language. The amount of diminution of the patient's vocabulary, the degree of inappropriate usage of words, the imperfections of sequence and rhythm, should all be noted. The patient should be tested for his power of recognition of simple words, short sentences, and long sentences. As he may react to conventional questions, uncommon requests should be made. The ability of the patient to interpret sounds should then be noted. Do sounds evoke previous memories of similar sounds, and do they incite the auditory centre to revive the name of the object from which sounds proceed? When a bell is sounded, or a watch is held behind the ear and apart from the stimulation of any perceptual avenue other than hearing, can the patient say "bell" or "watch?"

Then an attempt should be made to determine the various degrees of indentification of sounds and words which the patient hears, his capacity for repetition, and his capacity for spontaneous thought and action. His capacity to write spontaneously and from dictation, to read to himself, should then be tested, and then his ability to name objects and to designate persons and things.

Finally, the existence of any disturbance of bone or aerial conductivity should be demonstrated or excluded.

If word deafness can be excluded and the patient still makes no reply—that is, if he remains completely speechless—the examination should be made to determine whether or not internal language is defective, for the question has then narrowed itself to a determination of whether or not the aphasia is Broca's aphasia, or whether it is subcortical motor

aphasia. In other words, is the inability to speak due to a lesion of Broca's area, or is it due to lesion of the neurons that conduct the motor word impulses from the Rolandic area to the parts that externalize the word? The essential thing, then, is to determine if the patient is in full possession of the internal language. If the internal language in any of its components is disordered, then the patient has true cortical motor aphasia. If, on the other hand, there is no such disturbance, the lesion is elsewhere than in the zone of language. In some patients the differentiation will be an easy one. On the other hand, however, the task is oftentimes extremely difficult. It is particularly so because the test to determine if the legitimate idea of words can be evoked in the internal language (the test of Proust and of Lichtheim) is not one of universal application, because in the first place many patients have no sufficient scholarship to know anything of syllables or word construction. In the second place, there is very often associated with aphasia a slight degree of dementia. In such patients it is often extremely difficult to make them understand just what is meant by telling them to press the physician's hand as many times as there are syllables in a word. Nor is the substitute of asking the patient to make voluntary expiratory effort as many times as there are syllables or letters in a word more applicable. But even when we cannot get the patient to respond to these tests, there is a general atmosphere about the patient with subcortical motor aphasia (aphemia, anarthria) which enables one to recognize that he is in full possession of his intellect and internal speech. The only shortcoming of the subcortical motor aphasic is inability to articulate. He understands everything that is said to him; he interprets information received through the visual sphere; he is capable of expressing his thoughts fully, easily, and correctly by writing and by pantomime, or, at least, he would be if it were not that the right half of the body is usually paralyzed, and he is obliged to portray mental states by the pantomime activity of the left, the less dextrous half of the body.

Difficulty is often found in properly assigning cases of cortical motor aphasia, because the patient is still able to articulate some words. If it be kept in mind that the patient with Broca's aphasia need not be absolutely deprived of the power to articulate words; that he frequently retains the ability to say one or several words, which he uses at all times and under all conditions, and that frequently these words take the form of recurring utterances; that there is always agraphia, which is usually proportionate to the aphasia; that it is manifest in voluntary writing and in writing from dictation, but not in writing from copy, and that the patient in copying, copies print in script and script in print, showing that the copying is not a mechanical, but an intellectual act; and that there is defective internal speech, then the diagnosis of Broca's aphasia will not be so difficult.

After having tested the patient's capacity to perceive and interpret words through the auditory apparatus, he should be examined with a view to determining if there is any disability of acquiring and interpreting information through the visual apparatus. To do this requires patience and circumspection. In the first place, it should be established



that the patient has no trouble with the peripheral apparatus. This can be done by an ophthalmoscopic examination. Tests should be made to determine the existence of hemianopsia. This is not an easy matter to do if the patient is aphemic or if he has word deafness; in fact it is extremely difficult to do satisfactorily. With a patient who can understand what is said to him and who can indicate when he perceives the entrance of an object into the visual field, who can tell when the indicator of a perimeter passes beyond the range of vision, testing for hemianopsia is a simple matter. If the patient is word deaf, and if he has visual blindness, which, of course, he is apt to have if he has hemianopsia, one finds himself unable to convey to the patient by written or spoken word that which one wishes him to do or to observe. In such cases one must content himself with the information that is to be derived from forcibly and suddenly thrusting some object into the visual fields, from the right side (for right-handed patients invariably have right lateral homonymous hemianopsia when they have any), and taking note whether or not the patient blinks, as he should do if the object be perceived. If he does not, it is rather certain that he has hemianopsia.

To determine the integrity of the visual mechanism, one may show him familiar objects. If, having normal ocular apparatus, he does not recognize them, or shows by act or deed that he comprehends their uses or purposes, the lesion is probably of the occipital cortex. Such an individual may obtain information through the medium of other special senses, such as the tactile, gustatory, etc., that will enable him to recognize the object, the person, or the thing. If he is shown familiar objects, and he recognizes them, knows what they are for, but cannot name them, then he may have either an interruption in the pathway leading to the higher visual centre or in the centre of the intellect specialized for visual language. If it be the former, internal language will be preserved and spontaneous speech may be intact, although there is usually some paraphasia and jargon aphasia, and this preservation is shown most conclusively by the retention of ability to write. He may write easily and moderately well, not only voluntarily, but from dictation, but is unable to read what he writes. If the aphasia be of the latter character and complete, the patient will be absolutely agraphic. This agraphia is to be considered a part of the disorder of internal language; there is inability to arouse the visual image of the word. In such a case, an arousal must precede the transmission to the part of the Rolandic cortex that innervates the member holding the pen.

We shall understand aphasia better after we have interpreted the significance of apraxia, and in every case of aphasia the existence or absence of apraxia should be established from examination of the patient's spontaneity and initiative and by a study of movements conditioned by visual, auditory, tactile, and kinesthetic stimuli. It should be specifically noted what the patient does when objects are held up before him, what his responses are, and how they are expressed to appeals of any nature made to him through his vision, his hearing, or his senses of touch and posture. Apraxia may exist without aphasia, and vice versa, and when they occur separately the interpretation of each is much easier.

## CHAPTER IX.

### TUMORS OF THE BRAIN AND MENINGES.

By HARVEY CUSHING, M.D.

**Introductory.**—Under the caption “Brain Tumors” it is customary to group all neoplasms which encroach upon the intracranial space. In this broad sense the variety of new-growths which symptomatically compromise the brain, whether by displacement from pressure or by actual destruction, is multitudinous. They include not only the tumors originating from the tissue components of the brain and its various appendages, but those arising from the meningeal envelopes and even from the cranium as well; they include metastatic growths whose tissue elements may be foreign to the brain, and also certain infectious lesions—cysts and granulomata—which elicit the familiar pressure symptoms of tumor, though, strictly speaking, they are not neoplasms.

The characteristic symptom complex of an intracranial tumor may therefore be produced by lesions which originate from a cranial or extracranial source and implicate the brain secondarily through metastasis or invasion; although lesions of this type are numerous, for the sake of brevity they will be given only passing consideration. One must distinguish clearly between the growths which primarily occur within the substance of the brain and those which arise from its appendages or meningeal coverings.<sup>1</sup>

**Incidence.**—Despite their supposed rarity, the brain is actually one of the most common seats of tumor growth—their seat of predilection according to Oppenheim; and Bruns affirms that when a general practitioner says he has never encountered an instance it is an acknowledgment that patients with tumor have passed through his hands unrecognized. Though during the past decade physicians have largely awakened to this fact, it is still true in all probability that by far the larger number are incorrectly diagnosed unless at a terminal stage of the disorder. Furthermore, as the progress of the disease is often a lamentably slow one, it is with comparative infrequency that patients thus afflicted end their days in a general hospital. Hence it is unusual for them to figure largely in pathological records. In certain hospitals large numbers of these cases have been observed. Thus in the National Hospital, London, there were 500 cases between 1902 and 1911 (Tooth).<sup>2</sup> The writer's series comprises almost an equal number.<sup>3</sup>

<sup>1</sup> From the enormous literature there are a few comprehensive monographs which should be consulted: L. Bruns, *Die Geschwülste des Nervensystems*, 2te Auflage; S. Karger, Berlin, 1908; H. Oppenheim, *Die Geschwülste des Gehirns*, *Nothnagel's Spec. Path. u. Ther.*, Wien, 1896, Bd. ix; H. Duret, *Les Tumeurs de l'Encéphale*, Félix Alcan, Paris, 1905; E. Poggio, *Diagnostic di Sode dei Tumori Intracranin.* Unione Tipografica Editrice di Torino.

<sup>2</sup> *Brain*, 1912, xxxv, 64-108.      <sup>3</sup> *Journal of the Amer. Med. Assoc.*, 1915, lxiv, 189

In the autopsy reports of the Johns Hopkins Hospital to January 1, 1909, among 3150 autopsies there were 55 cases of brain tumor (1.7 per cent.). This is a larger percentage than that given by Siedel, who found, in Munich, 1.25 per cent., and by v. Beck, who found, in Heidelberg, 0.8 per cent. There are many elements to be considered, such as the average age of hospital patients, the character of the service, and the interest paid to neurological cases. Bruns states that 2 per cent. of all patients classified as neurological suffer from brain tumor, and Blackburn's figures (29 tumors in 1642 autopsies, nearly 2 per cent.) show an equal percentage for asylum patients.

In the medical wards of the Johns Hopkins Hospital, in the first 25,000 admissions, there were in the neighborhood of 100 cases diagnosed as brain tumor, making an average of one tumor in every 250 patients. In the surgical service during the same period the number of patients with the diagnosis of brain tumor rose from 0.06 per cent. in the first 5000 admissions, to 0.2 per cent. in the second 5000, to 0.3 per cent. in the third 5000, to 0.75 per cent. in the fourth 5000, and to 1.3 per cent. in the following 3000 cases. In the subsequent 1000 surgical admissions there were 40 tumor cases, showing the rapid increase when particular attention is given to them. This is further emphasized by the experience at the Peter Bent Brigham Hospital where in the first 2500 surgical admissions there were (including cases readmitted) approximately 200 cases (8 per cent.) of brain tumor or presumed brain tumor; this number is exclusive of the growths associated with the pituitary body.

**Etiology.—Sex.**—There is a general opinion that tumors occur with greater frequency in the male than in the female—twice as often, according to the following figures: Bruns found, in 63 autopsy cases, 40 males and 23 females; Gowers, in 650 clinical cases, found 440 males and 210 females. An analysis of the author's first 130 patients with tumor or presumed tumor showed 87 males and only 43 females.

**Age.**—More than half the cases occur between the ages of twenty and forty years. Tumors are said to be rare at the extremes of life, if tubercles and congenital growths of infancy be excluded. This, however, must be taken with a certain reservation, for in the 22 cases in the first decade, there was one dermoid cyst and one tubercle, the others being mostly cystic gliomas. Gowers limits the relative immunity of infancy to the first five months. Grouping the aforementioned 130 cases according to decades, there were 22 in the first, 13 in the second, 28 in the third, 37 in the fourth, 20 in the fifth, 8 in the sixth, and 2 in the seventh decade.

**Trauma.**—The relation between trauma and the first appearance of symptoms is a coincidence that occurs too often to be ignored, though it is possible that blows on the head may serve merely to bring a preëxisting lesion into symptomatic prominence; for, in consequence of the blow, the rupture of a bloodvessel may occur in a vascular glioma or concussion œdema may accentuate symptoms which were previously vague. The point of injury may even indicate the situation of the lesion and this would appear to be true even of the meningeal endotheliomata. It is more often the case, however, with the granulomata, for here we are dealing with a simple infection at a spot of lessened tissue resistance.

Indeed, the latent tumor itself, through an epileptic attack or faint, may have been the cause of the fall producing the cranial injury. Thus trauma may not actually occasion, but may only serve to bring into the open, the symptoms of a previously obscure growth. This is a debated question, and naturally one of the arguments advanced in its favor is the greater frequency of tumors in the adult male, whose occupations and sports engender blows on the head. Shaving the scalp preparatory to operation often reveals an unsuspected and significant scar.

**Pathology.**—The comments on the frequency of brain tumors indicate how misleading collective evidence may be; and this applies even more to the past statistics relative to their histological nature. Few pathologists have had opportunity or inclination to make a comprehensive study of a large series of cases, for a complete postmortem investigation entails three factors: (1) The gross relations and physical character of the tumor in so far as it disturbs the brain in general as well as its circulation; (2) the histological nature of the lesion itself; and (3) the secondary degenerations. The first requires the careful fixation of the tissues *in situ* by carotid injections—a precaution which is rarely taken. The second necessitates absolutely fresh tissues, for the postmortem changes which occur rapidly in the central nervous system militate against the perfect fixation essential for fibrillar stains. The third object of study—the degenerations—can only be of contributory value in exceptional cases when lesions are small, uncomplicated, situated in such areas as to have involved paths whose degenerations are profitable for study, and finally when the pathological findings can be correlated with an exhaustive clinical record. One or another of these is usually lacking.

Mallory's studies<sup>1</sup> have shown that it may be impossible to make positive statements in regard to the cellular structure of many tumors, particularly of those arising in the nervous system, without operative removal and immediate tissue fixation. This goes to show with what caution one must accept earlier statistics on the frequency of glioma and sarcoma. A matter of equal importance has been emphasized by Adolf Meyer and Charles J. Lambert, namely, the desirability of a histological study of the entire growth *in situ*. The section of an entire growth will oftentimes betray its nature, owing to a certain characteristic arrangement of the cells which cannot be appreciated by the survey of a small fragment. This applies particularly to the recognition of certain gliomata.

The histological diagnosis in our early (Baltimore) series of 130 cases was authenticated both by Dr. Mallory and by Dr. Lambert, so that with the exception of one or two tumors of very obscure nature we may speak of them with some certainty. The tissues became available either through operation or autopsy in 69 cases. Of these, 44, or 66.6 per cent., proved to be *gliomata* or gliomatous cysts (11 cases, in 3 of which a demonstrable shell of tumor was still present), or gliopsammomata (3 cases). Many of these in the past would have been classed with sarcomata, of which there was but a single instance (metastatic) if we exclude the 5 endotheliomas. There were 4 tubercles and 3 syphilomas: with the rest of the

<sup>1</sup> *Jour. of Med. Research*, 1905, viii, N. S., p. 113.

field scattering. In Allen Starr's much quoted table,<sup>1</sup> based on 600 cases gathered from various sources, at a time when histological differentiation was impossible, there were 91 gliomata, only 15 per cent.; whereas there were 120 cases, or 20 per cent., tabulated as sarcoma. Hence sarcoma was long regarded as by far the commonest tumor in the adult. In 70 certified lesions in our more recent (Boston) series the diagnosis was as follows: glioma and gliomatous cyst, 27 (38.5 per cent.); endothelioma, 7; angioma, 2; sarcoma, 1; carcinoma metastatic, 1; syphiloma, 4; tuberculoma, 4; osteoma, 1; echinococcus cyst, 1; adenoma, 17 (hypophyseal 15, pineal 2); interpeduncular and mixed tumors, 5. Excluding the pituitary cases the gliomas would represent about 50 per cent. of the series. In Tooth's studies<sup>2</sup> of the cases at the National Hospital, in 258 certified cases 127, or 49.2 per cent., were gliomas. So that one may say that one-half of all brain tumors are gliomas of one kind or another.

**Gliomata.**—These were first shown by Virchow to represent specific growths of the neuroglial tissue framework of the central nervous system; and since the discovery of a specific stain for neuroglial and ependymal cells, Mallory<sup>3</sup> and Stroebe have demonstrated that they may arise from the ependyma as well. A glioma consequently may arise in remote situations, as the coccygeal region, from vestiges of the primitive neural canal. Christian has shown that ependymal epithelium and neuroglia are common constituents of teratoid tumors of ovarian origin. Thus, a glioma is a growth of ectodermal origin, whereas a sarcoma arises invariably from the mesenchymal tissues. A true combination of these forms is rare, although the difficulty of distinguishing them without special stains and close study has led to the misuse of the term "gliosarcoma."

Cells are found in gliomata showing all gradations from the undifferentiated embryonal type to the characteristic neuroglial spider cells on the one hand and toward ganglion cells on the other. These growths doubtless have an embryonic origin, and thus a single cellular *anlage* suffices to explain all the types of cells which are found—neuroglial, ependymal, or ganglionic. The growth may be made up almost entirely of round, undifferentiated cells, possessing no fibrillæ, so that only by some characteristic arrangement of these cells, or by the finding of a small group which has begun to assume a more adult type, can such a tumor be distinguished from certain forms of sarcoma. Lambert is of the opinion that there is a fairly uniform tendency for the cells to arrange themselves more or less radially around a central core, which, on cross-section shows a characteristic rosette-shaped figure—the satisfactory disclosure of which may require a section through the entire growth.

The variable differentiation of the primitive cells into those of a ganglionic type or into ependymal cells has led to the terms *neuroglioma ganglionare*, *ependymal glioma*, etc. The *neuroglioma* of Klebs, in which the nerve fibres themselves participate in the new-growth, becomes a

<sup>1</sup> *Brain Surgery*, Wm. Wood & Co., N. Y., 1893, p. 202.

<sup>2</sup> *Brain*, 1912-13, xxxv, 61-108.

<sup>3</sup> *Jour. of Med. Research*, 1902, iii, N. S., p. 1.

somewhat doubtful lesion in view of the known persistence of normal preëxisting fibres which traverse many of these tumors.

A glioma is usually a solitary tumor which may occur in any part of the cerebrum, cerebellum, or brain stem. There is often an extensive gliosis which spreads widely over the nervous system with but a single area which in gross is evidently a neoplasm. In a few of the primary cerebellar gliomas in the author's series the gliosis had in time extended throughout the length of the entire spinal cord. Though such a gliosis is a diffuse process the tumor spreads by extension and in a strict sense does not form metastases, although regionary extensions which resemble them may occur. They may be of cortical or basal origin, or may arise, although less frequently, from the ependyma. Gliomata supposedly originating in the corona radiata are common and may attain a large size. The *ependymal gliomata* which arise from the subependymal layer of the ventricles, possess unusual characteristics. Ten out of the twelve published cases, according to Martens and Seiffer,<sup>1</sup> originated, as did their own, in the fourth ventricle, which seems to be their seat of predilection. The striking feature of these growths is their multiple nature, for they have a tendency to spread, so that small, metastatic nodules may be found throughout the ventricles, resulting, in rare cases, in an extensive diffuse growth—*ependymgliomatosis*. Their symptomatology is obscure and they seem never to have been diagnosed clinically.<sup>2</sup> There have been two cases in the author's series in both of which the tumor had extended down on the dorsum of the spinal cord.

In color, gliomata resemble the grayish red of cortical tissue, and their consistence is not unlike the normal brain, so that in the absence of pigmentation from hemorrhages, from cystic or other structural metamorphoses, it may be difficult to distinguish them from the surrounding tissues when exposed during an operation or even at autopsy. This is particularly true of certain forms of *diffuse gliomatosis*, often mistaken (Gowers) for *hypertrophies* of the brain. Some of the largest recorded intracranial tumors have been gliomata. A gliopsammoma in our series weighed 496 grams.

Gliomata are usually described as infiltrating tumors with an indefinite boundary which shades off into the normal tissues without clear demarcation; but this is not always the case, for they may be encapsulated. By their character of growth it frequently occurs that the form of the brain remains unchanged, with complete absence of pressure symptoms, and also that normal functioning nerve fibres are found traversing them.<sup>3</sup> This explains the latency of many gliomatous growths and also shows how their extirpation may greatly accentuate focal symptoms which were slight before their operative removal. The pia usually serves to check their extension, although this is not invariable, for we have seen a number of gliomata which have broken through into the subarachnoid spaces and spread over the surface in the manner of an infective process.

Gliomata are often exceedingly vascular (*gliomateleangiectodes*), and

<sup>1</sup> *Berl. klin. Woch.*, 1908, xlv, 1477.

<sup>2</sup> Bassoë, *Archives, of Int. Med.*, 1908, ii, 194.

<sup>3</sup> Byrnes, *Jour. of Nerv. and Ment. Dis.*, 1909, xxxvi, 129.

trauma, congestion, or operation with dislocation of position, may lead to hemorrhages from rupture of the thin-walled vessels. A spontaneous hemorrhage simulating apoplexy may occur and be the first symptom of an unsuspected growth; or repeated small hemorrhages may occur, with marked fluctuation of symptoms. Degenerative processes with cyst formation (*cystoglioma*) are common, and the entire growth may seemingly become replaced by a single cyst, although a careful search will often reveal vestiges of the original growth on the cyst wall. Areas of calcification—*gliopsammoma*—or even ossification, may occur.

Certain tumors of mesenchymal origin (sarcomata) can only be distinguished from some of the gliomata by stains of absolutely fresh and properly prepared tissue. Gliomata were claimed by Ströbe<sup>1</sup> to be distinguishable from sarcomata by their gross appearance and manner of growth, for the latter are encapsulated, are sharply distinguishable from the brain, and easily enucleated. This distinction is not to be relied upon, for a number of gliomata in our series have shown all the features, except histological ones, thought to characterize sarcoma.

**Endotheliomata.**—Next to the gliomata these tumors, often misnamed psammoma, sarcoma or fibroma are probably the most frequent of all true intracranial tumors (14.3 per cent., Tooth; 11.1 per cent., Cushing), though they take their origin from the endothelial lining of the meninges much more often than from an endothelial source, vascular or lymphatic, within the substance of the brain itself. Hence, they are apt to be extracerebral or extracerebellar, as the case may be, rather than intracerebral or cerebellar, and in a strict sense are not brain tumors. They are commonly called *dural endotheliomas*, but it is the author's belief that they arise from the clusters of mesothelial cells of the arachnoid villi or tufts which project into the dura and which are the points of outlet for the cerebrospinal fluid.<sup>2</sup>

In the statistical tables of the past they have been included with sarcomata or fibromata, for their spindle-shaped cells resemble those characterizing these growths. Presumably all of the so-called benign fibrosarcomata originating either from the cerebral or spinal meninges have been of this nature, and the diagnosis of fibroma of the brain is open to doubt. Mallory has demonstrated that fibrils do not occur in these tumors, so that they can be thus differentiated histologically from fibromata or sarcomata.

These tumors are intrinsically benign, but ultimately cause pressure symptoms by slow indentation or moulding of the cerebral substance. They do not invade the cerebral tissues, but can be readily dislocated from their cavity or nest, leaving a smooth surface. They grow slowly and are often of long duration. In two of our cases the lesion had doubtless been present, giving local symptoms, for thirteen and nine years respectively before pressure disturbances of serious import, due in each to an obstructive hydrocephalus, became pronounced. Blackburn's figures indicate that a considerable proportion of the asylum cases showing tumor after death have this lesion.

<sup>1</sup> *Ziegler's Beiträge*, 1895, xviii, 405.

<sup>2</sup> Studies on the Cerebrospinal Fluid, Cushing and Weed, *Jour. Med. Research*, 1914.

Endotheliomata may attain a large size (one tumor in the author's series weighed 246 grams on removal) and they may occur in almost any situation over the hemisphere. They, however, have certain seats of predilection, a very common source of origin being from the arachnoid villi along the sagittal sinus. They not infrequently cause an osteoma-like dome-shaped thickening of the overlying skull. They may, on the other hand, cause a pressure absorption of the adjacent skull. Another supposedly common seat of these tumors—possibly due to the unmistakable symptoms elicited—is the cerebellopontine recess. Occasionally bilateral tumors may be found in this situation. Owing to their presumed origin from the sheath of the acoustic and the early onset of auditory symptoms, they are often spoken of as *acoustic fibromata*. They are, however, to be distinguished from the intracranial *neurofibromata* of von Recklinghausen's disease, which are apt to be symmetrical and may be multiple, involving other nerves than the auditory. Endotheliomata sometimes undergo degenerative processes, and large gelatinous or cystic areas may be found. Of all tumors, they are the most favorable for operative removal.

**Sarcomata.**—The so-called gliosarcomata are in all probability gliomata made up in part of undifferentiated cells; the fibrosarcoma of the past is, in truth, a meningeal endothelioma. Still, if one is to include all actual sarcomata which serve directly or indirectly to implicate the encephalon, they may perhaps continue to represent a form of cerebral growth which is common enough, even though they no longer out-number other tumors. They are nowise specific of the nervous system, and although they may originate there, are more often invasive or metastatic in origin. They may be intracranial or extracranial growths.

Although varying greatly in consistency, being in general of a firmer structure than most gliomata, they nevertheless are sharply distinguishable from the brain tissue in which they lie more or less definitely encapsulated and from which they are easily shelled out. There is usually a zone of softening of the surrounding brain due to vascular disturbances. This would make them favorable for surgical removal were it not counter-balanced by the frequent presence of secondary metastatic nodules and the tendency to involve adjacent tissues.

These tumors, often of cranial origin, include the *alveolar sarcomata*, growths which, without fibrillar stains, it may be impossible to distinguish from endotheliomata or from true epithelial tumors. They are often of diploëtic origin, the temporal bone constituting their favorite seat (Weisswanger). *Osteosarcomata* likewise take their origin from either the cranial vault or base and, penetrating the skull, lead to pressure symptoms. Endotheliomata arising from the meninges may also come to involve the bone and assume certain outward characteristics of an osteosarcoma from which they must be carefully differentiated. These so-called *perforating sarcomata* may arise from either surface of the dura, from diploë or either table of the skull, and as they infiltrate the normal tissues and even absorb the bone in their process of growth, they are apt in time to give external evidence of their situation. As the bony absorption takes place slowly, the growths are apt to assume an hour-glass shape, the



intracranial portion of the tumor indenting, but usually not invading, the cerebral cortex, which remains protected by pia-arachnoid. When the scalp has become broken down the condition known as *fungus duræ matris* results. The brain is occasionally involved secondary to a cranial lesion by the growths designated as *chloroma* or *myeloma*.

Sarcomata found within the substance of the brain are usually of metastatic origin and show the character of the original tumor. They may be cystic, or may undergo cystic degeneration (*cystosarcomata*)—a process which occurs possibly less often than is the case with gliomata. They also may become oedematous and the cells assume a mucoid character—*myxosarcomata*. Other degenerative processes may lead to necrosis or caseation, when they may bear a close resemblance to tubercle. Deposits of calcification may occur within them, giving rise to *psammosarcomata* or angiolithic sarcomata. Intracranial growths with psammomatous bodies, whether gliomatous or sarcomatous, can occasionally be localized by the x-rays. *Pigment-containing sarcomata* are said to occur in the brain only in metastatic form and the metastases, as a rule, are numerous. The single indubitable sarcoma in our series was a large, coal-black, encapsulated melanosarcoma removed at operation from a subcortical situation in the parietal lobe. At the time there was no obvious extra-cerebral source of origin. The patient died a year later, and a primary melanotic growth of the ovary was found with numerous small, visceral metastases and two other nodules occupying silent areas in the brain.

**DIFFUSE SARCOMA.**—There is a particular form of diffuse sarcoma (pachymeningitis interna sarcomatosa, Orth) in which a primary sarcomatous thickening of the meninges may occur almost over the entire central nervous system—brain, cord, and nerves often being encased by the nodular masses; it would seem as though a direct implantation of tumor cells had occurred through the medium of the cerebrospinal fluid. Occasionally a primary growth seems to have burst into one of the lateral ventricles, leading to the inoculation, as it were, of the cerebrospinal fluid with wide dissemination of the infective agency. The process in less extensive form is more commonly met with about the base of the brain, often with bilateral involvement of the cerebral nerves, and not infrequently of the cauda equina as well.

**Carcinomata.**—Like the foregoing these tumors occur for the most part as metastases, being found most often as secondary nodules in cases of mammary cancer. The nodules occur usually in the substance of the brain, are frequently multiple, and show a marked tendency to cystic degeneration. Epithelial tumors occasionally arise from a cranial or extracranial source and invade the cranial chamber secondarily. We have seen such a tumor apparently of labyrinthine origin.

**Epitheliomata and Papilloma.**—Primary epithelial tumors may arise in the brain from the secreting cells of the choroid plexus (Ziegler), and one of the tumors in our series, difficult to classify, is a large, seemingly benign epithelial growth arising in the occipital ventricle and containing cysts with gelatinous contents, apparently of this origin. These tumors are regarded by some as a simple hyperplasia or *adenomatous papilloma*

of the plexus; 38 analogous cases have been collected from the literature.<sup>1</sup> Epithelioma of the ependyma has been described, and the hypophysis is naturally a not infrequent seat of epithelial as of other growths. These are commonly a form of malignant adenoma, into which a pituitary struma has become transformed much as a thyroid struma may undergo malignant changes.

**Cholesteatomata.**—Sometimes called “mother-of-pearl” tumors in consequence of their peculiar glistening external appearance, these growths have their seat of predilection in the basal meninges. They apparently never arise primarily in the brain substance itself, although this becomes excavated and compressed as the growth advances. There is no unanimity of opinion as to their histogenesis. Virchow, Frank, and others thought that the cells of the arachnoidal meshwork multiply and distend the pre-formed endothelial spaces, producing a sort of complex retention cyst rather than a true tumor growth. A later view, advanced by Ziegler, Bostroem, and others, favors a congenital *anlage* of misplaced epidermoidal cells, supported by the fact that the cells become cornified and contain keratohyalin. Bostroem has demonstrated that a close relationship exists between these and the more usual forms of dermoid tumors. Possibly both epithelial and endothelial forms may exist.

The otitic cholesteatoma of older writers, following chronic suppurations of the middle ear, apparently consists of epidermoidal masses which cause pressure absorption of the temporal bone and invade the cranial chamber secondarily. These tumors usually possess a more or less irregular flat form, modified as the growth pushes its way into the clefts and recesses between the bone and base of the brain. The surface is often covered with warty irregularities. On section these tumors consist of a grayish, friable, more or less laminated mass, made up of layers of a closely packed mosaic of flat, polygonal cells—really a dead tissue containing no bloodvessels. They are of slow growth and may run a symptomless course or give definite general and focal symptoms of a basilar tumor with cerebellar, cerebellopontine, or trigeminal symptoms.

**Dermoid Tumors.**—These are among the rarest of intracranial growths. There is a single instance in our series, the cyst, containing hair, having been successfully extirpated. Bostroem considers that various grades are recognizable, from a simple epithelial-lined sac, which he would regard as a cholesteatoma, up to the most complex embryoma which may contain examples of many tissues of the body. These tumors date from the third to the fifth week of embryonic life. They are usually basal, commonly mesial, grow slowly, and are not likely to produce symptoms. Another variety is connected with the formation of dura and bone, and it is not uncommon for them to connect, through a cranial defect, by a stalk with an extracranial dermoid.

There are other lesions, possibly somewhat less rare than the foregoing, which deserve passing mention to complete the enumeration.

**Teratomata.**—These have been found, chiefly in the neighborhood of the interpeduncular space, arising doubtless from some embryonal

<sup>1</sup> Boudet et Clunet, *Arch. de Méd. Expériment et d'Anat. Patholog.*, 1910, xxii, 379.

vestige with faulty development of the pituitary gland. They are apt to be accompanied by symptoms of hypopituitarism and by the characteristic local symptoms of tumor in this situation.<sup>1</sup>

**Chordomata.**—These tumors arise from an *anlage* at the cephalic end of the primitive notochord. They are usually small but may undergo malignant transformation and reach a large size as in the case recorded by Jelliffe and Larkin.<sup>2</sup> They are situated in the interpeduncular space and the resemblance to cartilage possibly leads to a diagnosis of chondroma or teratoma, and this may account for their presumed rarity.

**Lipomata.**—These are rare and those described have usually been small, pea-sized, basal growths in which bone is often present. Zuckermann has collected 50 such cases. A larger form is known which has especial predilection for the upper surface of the corpus callosum. Hecht has described a lipoma of the Sylvian Fissure.

**Fibromata.**—These are occasionally found in fairly pure form, arising from the periosteum of the cranial base or from the nerve sheaths—the latter being more properly classified as a manifestation of von Recklinghausen's disease. It is presumable that the endothelial cells of an endothelioma may become fibroblastic, and thus many so-called fibromata really belong to the endothelioma group.

**Neuromata.**—The same is true of the so-called neuromata, which arise from the perineurium of the cerebral nerves, and consequently are extracerebral tumors. They usually surround the entire nerve, giving it a spindle-shaped form, and may undergo sarcomatous or myxomatous change. They are particularly common on the acousticus, but other nerves may be involved (multiple neurofibromatosis).

**Neurocytoma or Neuroblastoma.**—These terms have been introduced by J. H. Wright<sup>3</sup> to cover a group of tumors the essential constituents of which are undifferentiated nerve cells (neurocytes) and the fibrils of which resemble those occurring in the *anlage* of the sympathetic nervous system and chromaffin tissues. There may be a simultaneous involvement of adrenal, liver, and brain. The single example identified in our series had been regarded as a glioma.

**Enchondromata.**—These have been found arising from the choroid plexus or meninges. One especial variety, arising from the cartilaginous vestige of the *Clivus Blumenbachii* (Virchow), has its seat of predilection near the pituitary fossa (cf. chordoma and teratoma).

**Angiomata.**—Angiomata, whether simple, telangiectatic, or cavernous, are congenital and usually cortical. As a rule, they produce no serious symptoms, barring the possible accident of hemorrhage or thrombosis. Typical pressure symptoms were elicited in one of our cases by a large, cavernous angioma of the temporal region. The *angioma racemosum* may occur alone or with congenital vascular lesions of the scalp or face (Kalischer; Emanuel; Cushing). Cavernous angiomata are rare.<sup>4</sup>

<sup>1</sup> H. Cushing, *Jour. of Nerv. and Ment. Dis.*, 1906, xxxiii, 704; D'Orsey Hecht, *Jour. Am. Med. Assn.*, 1909, liii.

<sup>2</sup> *Jour. of Nerv. and Ment. Dis.*, 1912, xxxix, 1.

<sup>3</sup> *Jour. of Exper. Med.*, 1910, xii, 556.

<sup>4</sup> Astwazaturoff, *Frankf. Zeitschr. f. Pathol.*, 1910, iv, 482.

**Psammoma.**—This is a term given to a number of tumors, glioma, sarcoma, and endothelioma, in which areas of calcification have taken place, making them gritty on section. Bruns is inclined to restrict the term to certain growths which arise in the pineal gland or choroid plexus where, normally, calcareous bodies—the so-called “brain sand”—occur.

**Cysts.**—We meet with (1) congenital interpeduncular (pituitary) cysts; (2) simple serous cysts which presumably are degenerated tumors; (3) tumors containing cysts, as cystosarcoma, cystoglioma, etc., possibly on the way to the above; (4) blood cysts, the result of cerebral or cerebellar apoplexy; (5) cysts the result of softening after embolism; (6) the cystic widenings or diverticula of the fourth ventricle; (7) dermoid cysts; (8) parasitic cysts; and (9) gas cysts.

The congenital interpeduncular or suprasellar cysts, the *Hypophysen-erganggeschwülste* of Erdheim, arise from a pharyngeal *anlage* associated with the development of the pituitary body. The lesion is not an uncommon one and is usually, although not always, associated with evidences of pituitary insufficiency together with optic atrophy with bitemporal blindness. Characteristic outgrowths of squamous (pharyngeal) epithelium arise from the walls of these cysts.

It is probable that, in this part of the world at least, the greater number of so-called *simple cysts* are actually degenerated tumors (gliomata and sarcomata), although it may be difficult to demonstrate this conclusively. Since surgical explorations have become frequent, it has often been found that tumors originally solid have become cystic at a later exploration or at autopsy; and one occasionally sees a tumor in what seems to be a process of cyst formation with a large area of semifluid, degenerated tissue.

Unlike those which may form as the result of absorption of an apoplectic extravasation, and also unlike the porencephalic cysts of childhood, these degenerative cysts may continue to give pressure symptoms almost as severe as those of the original growth. On evacuation they tend to refill. Their fluid content is often of a yellowish or brownish tinge, and the fluid, rich in fibrin, clots on exposure to the air. Their seat of predilection is the cerebellum. Scholz<sup>1</sup> has made an elaborate study of 75 published cases. There have been many cysts in our series, the larger percentage of them cerebellar; and we have seen a number of borderline conditions in which a glioma seemed to be in the process of total cystic degeneration after a decompressive operation.

There are certain obscure conditions, often seen in association with an actual tumor growth, in which a large cyst of the hemisphere has an apparent communication with a lateral ventricle.

**Arachnoidal Cysts.**—These cysts, a consequence of trauma, may give pressure symptoms and focal signs simulating tumor. They may follow lesions which leave a defect in bone and dura over a given area, for an accumulation of the fluid which exudes through the arachnoidal membrane may be under tension, giving focal pressure symptoms.

**Parasitic Cysts.**—The *Cysticercus cellulosæ* shows an especial predilection for the central nervous system. Thus in 87 cases in Berlin the cysts

<sup>1</sup> *Mitteil. a. d. Grenzgeb. d. Med. u. Chir.*, 1906, xvi, 745.

in 72 instances were cerebral (Bruns). The hooked embryos attach themselves to the meninges, cortex, or ventricular wall—often of the fourth ventricle—where the larva develops as an isolated bladder-like structure, rarely exceeding a centimeter in diameter. The cysts are apt to be multiple, 111 of them having been counted by Delore and Bonhomme in a single patient. A particular form sometimes occurs in the basal meninges and a condition of so-called “cysticercus meningitis” may result. Ventricular cysts usually lead to a great increase of fluid and a distinct ependymitis may result (Lloyd).

The larvæ may be long-lived, although it is usual for them to shrink and become calcified in from three to six years. An especial study has been made by Stern<sup>1</sup> of the 68 known cases of fourth ventricle involvement. The recent presence of an adult *tænia solium* in the intestine should arouse suspicion of the nature of the case.

*Echinococcus* or *Hydatid Cysts*.—These may likewise occur in the central nervous system. The cysts are more apt to occur in the brain substance than in the meninges or ventricles. They are said to be usually single though in the only case in our series three were removed at operation from one hemisphere. They may reach a large size, and show an especial tendency to absorb the overlying skull and thus to appear under the scalp. Suppuration may occur, or the worm may die and the contents containing the free hooklets become gelatinous and thick. The symptoms are those of a slowly growing tumor; and on exposure a differential diagnosis from other forms of cyst can at times be assured only by the demonstration of head or hooklets, or by the peculiar structure of the cyst wall.

**Infectious Granulomata.**—Formations resembling tumors are the frequent result of tuberculous or syphilitic, and occasionally of actinomycotic, processes. Although not neoplasms in the same sense as some of the foregoing, custom calls for their consideration with tumors.

**ENCEPHALIC TUBERCULOMATA.**—Of encephalic tuberculomata two forms are recognized: (1) Focal *tuberculous meningo-encephalitis*, and (2) the so-called *solitary tubercle*. Tuberculosis of the central nervous system is usually—perhaps always—a complication of tuberculosis elsewhere.

*Focal Tuberculous Meningo-encephalitis.*—This form consists of a local plaque of disease made up of a conglomeration of more or less caseated tubercles, usually situated on the surface of one of the cerebral hemispheres, less often a cerebellar or a basilar process. Although of meningeal origin, more or less involvement of cortical tissue is inevitable, and with the advance of the process the characteristic focal symptoms of tumor may occur, although general pressure phenomena are usually absent until meningitis supervenes.

*Solitary Tubercle.*—The solitary tubercle behaves, on the other hand, in all respects like a brain tumor. According to Starr's tables, this should be the most common form of growth (32 per cent. if all ages are included), representing in the first two decades 50 per cent. of all cases, falling to 14 per cent. in adults. In Tooth's table tuberculoma occurred

<sup>1</sup> *Zeit. f. klin. Med.*, 1907, lxi, 64.

in only 5.4 per cent. of the cases and this corresponds closely with our own experience (5.7 per cent.). A tubercle has been found at the early age of twenty-three days in a case described by Demme.

The term "solitary" is somewhat misleading, for these individual growths are composed of many fused tubercles. "Solitary tuberculoma" is a better term, or "conglomerate tubercle," as Schmaus suggested. The lesions are multiple, at least at an early stage, in about 50 per cent. of all cases (Bruns), becoming fused later into an irregular conglomerate mass. This is the more likely since the originally discrete foci, in evidence of their embolic origin, are apt to be distributed on a single branch of the arterial tree. Their form is roughly spherical, unless modified by fusion or other especial influence. Their favorite seat seems to be in the cerebellum or brain stem. The individual lesions vary greatly in size, from a millet seed to a hen's egg on the average, although in rare cases by fusion of many isolated foci almost one entire hemisphere may become transformed into a tuberculous mass.

Coagulation necrosis with caseation takes place as the lesions enlarge, and on section the mass presents a dry, yellowish, crumbling centre, with a grayish-red, peripheral growing zone where are located viable bacilli and actively growing tubercles. As in tuberculous processes elsewhere central liquefaction with abscess formation may occur; less often the process dies out and calcification occurs. When the lesions are active the surrounding encephalic tissues are usually œdematous, and this tends to increase the pressure effects and constitutes a mechanical difference between a tuberculous and a sarcomatous tumor; for the latter, as a rule, only affects the immediately adjoining zone by pressure softening.

In no one of our several cases of extirpation of a cerebellar tuberculoma has the recovery been permanent. All have succumbed in the course of a few months to the effects of other lesions of like nature.

**SYPHILITIC PROCESSES.**—Syphilitic processes giving a tumor symptom complex occur in the central nervous system in two especial forms: (1) As a local spreading *meningo-encephalitis*, and (2) as a solitary granuloma—the so-called *gumma* or *sphiloma*. To these may be added the basilar *meningitis gummosa*, a process often arising in the middle cranial fossa where, with notable fluctuation of symptoms, it implicates one after another of the nerves which pass through the region. Vascular lesions are common, and sudden hemiplegia may occur from occlusion of the middle cerebral artery. This basilar process is apt to spread over the sylvian region, and, as the cortex usually is more or less involved, it hardly deserves to be distinguished from the more common *meningo-encephalitis syphilitica* of the convexity, except for its peculiar local manifestations.

Bruns, in agreement with Oppenheim and Gowers, regards syphilis as the most frequent of the conditions producing the tumor symptom complex in adults. This opinion is based naturally on the subsidence of symptoms after antisiphilic therapy—not an infallible test—and is contrary to postmortem statistics. It is doubtless on the same clinical basis that the cerebral cortex and the paracentral convolutions in particular are regarded as the favorite seat for gumma, owing to the definitely

localizing symptoms of the lesion when in this situation. The well-recognized association of these lesions with trauma is possibly a sufficient explanation for their frequent occurrence under the exposed vault.

*Syphiloma*.—Unlike tubercle, syphilitic brain lesions are largely confined to adult life, and are said to occur above the tentorium more often than below. In the majority of cases, furthermore, they are surface lesions, whereas tubercles are more common in the substance of the brain. For the most part, the solitary lesion does not reach a large size, and never equals the enormous, fused tuberculoma. Somewhat like an early tubercle, however, the gumma consists of a vascular, grayish-red, granulation tissue mass of round, epithelial, and giant cells. In this tissue mass changes soon occur; areas of coagulation necrosis appear, or through fibrosis a dense, scar-like tumor may be left—the typical syphiloma. This change is particularly common in the meningeal gummata.

In an advanced stage, cortex, meninges, and even overlying bone may become adherent in a dense cicatrix which resists absorption under the most vigorous antiluetic measures. Such a granuloma rarely undergoes the process of softening characteristic of the tuberculous lesions, although, on the other hand, disturbances of the surrounding cerebral tissue through accompanying vascular disease are much more common. Nevertheless, it is often difficult to distinguish the processes pathologically without microscopic demonstration of the agent—bacillus or spirochæte. One histological difference lies in the presence throughout the gumma of the bands of tissue undergoing fibrous change. We have occasionally experienced difficulty in distinguishing between a cortical gumma with spreading meningo-encephalitis and a superficially placed glioma which had invaded the pia-arachnoid. In all cases the serum reaction should be tried.

*ACTINOMYCOMATA*.—These are rare and have usually been secondary or metastatic lesions, although Bollinger regarded his cases as primary. Howard has made a thorough study of these conditions.<sup>1</sup>

**Pathological Physiology and Symptoms.**—The clinical aspects of a brain tumor hinge far more upon matters relating to the physiology of the intracranial space than upon its mere structural features. Indeed, in view of their extraordinary variability in size, form, consistence, manner of invasion, rapidity of growth and situation, not only in so far as the latter relates to the various nerve tracts and centres, but also to the blood supply and channels of outlet for the cerebrospinal fluid, it is perhaps remarkable that any recognizable symptom complex can be accepted as in the main characteristic of brain tumors.

There are, however, in the majority of cases, two processes whose phenomena may be regarded as typical of the greater number of intracranial new-growths. The first embraces the symptoms produced within an intact skull by a more or less generalized increase of tension which affects the brain chiefly by upsetting its normal vascular and cerebrospinal fluid circulations—the so-called underlying or *general symptoms*; and the second, the symptoms produced by irritation or

<sup>1</sup> W. T. Howard, *Jour. of Med. Research*, 1903, iv, 301.

destruction of nerve tracts or centres of known function—the so-called localizing or *focal symptoms*.

When both of these processes are in full operation—a thing which is to be expected eventually in most cases—the presence of the lesion, and probably its situation also, is readily recognized. Unfortunately, at the stage when the diagnosis may be of greatest value, one or both of these elements are wanting, and owing to the fragmentary sketch instead of the finished picture, the condition may long fail of recognition even by an expert. For example, the general pressure symptoms might naturally be expected to bear some relation to the size of the growth, and yet a small tumor compressing the aqueduct of Sylvius so as to obstruct the ventricular outflow may lead to symptoms out of all proportion to its size. On the other hand, a large infiltrating glioma may be present without eliciting general or focal symptoms; for it may invade the normal tissues in such a way as not to increase tension and it may be traversed by persisting fibres which can transmit impulses. Again, a small tumor too insignificant to produce pressure phenomena may, nevertheless, lead to definite focal symptoms, shown, for example, by irritative movements if it happens to be situated near the precentral gyrus; whereas a similar lesion over a silent area need give no indications whatsoever of its presence. Thus, the situation may be far more vital than the size.

It can be easily seen that tumors which happen to arise in a so-called silent area of the brain may remain long dormant, and that the first symptoms may be those of general pressure when, in the course of time, the lesion has considerably augmented in size. Thus patients may present themselves, with (1) tumors which give absolutely no recognizable evidence of their existence and are found post mortem; (2) tumors which present focal symptoms alone with no evidences of a general increase in pressure; (3) tumors which give general symptoms alone with no focal manifestations, when they happen to occupy a silent area; (4) tumors which give typical symptoms of general pressure together with definite focal symptoms; and (5) the symptom complex (general and local) of tumor, which in the absence of a growth is brought about most often by œdema from one source or another.

With full realization that practically all tumors at their onset fall under the first of the groups cited above, and also that many brain tumors may exist for years before their symptoms become sufficiently pronounced to attract any serious attention, we, nevertheless, may wisely retain as characteristic in general of most tumors at some stage of their progress the so-called underlying or general pressure symptoms—headache, vomiting, and choked disk—to which we would like to add the external evidences of intracranial venous stasis, the tendency to herniate through the cranial foramina or through operative openings and a few other possibly less characteristic signs of tension.

**The General Manifestations.**—That the general symptoms are primarily the consequence of pressure against the brain within the closed skull is now fairly conclusive, for they may be completely set aside by making a purposeful cranial opening, just as nature may occasionally alleviate them in the young by a general diastasis of the cranial sutures.



DISLOCATION FROM PRESSURE AND TENDENCY TO HERNIATE.—Physiological studies, particularly those of Leonard Hill, have shown that the brain itself is practically as incompressible as water, and consequently that any considerable encroachment by a new body on the intracranial space must greatly increase the cerebral tension through the medium of its fluid contents. One of the earliest consequences of this is an engorgement of the venous circulation, leading not only to the subjective sensation of fulness, but to certain objective signs. In its most familiar aspects this appears as a dilatation and tortuosity of the retinal vessels—the premonitory features of a choked disk. There are, however, other external evidences equally characteristic, although less often commented upon, which occur; for, as in the case of the retinal veins, the veins of the scalp, and particularly the smaller venules of the eyelids, often become a tell-tale of the intracranial venous stasis.

The cerebrospinal fluid is normally under low tension, and if there is no obstruction to its escape by usual channels the encroaching lesion at an early stage will drive the normally small amount of fluid out of the arachnoidal spaces, so that when exposed they will, relatively speaking, be found dry. Usually, however, the fluid becomes pent up in certain areas—in the interpeduncular arachnoidal space, for example, and hence the distension of the optic sheath; or in the ventricular cavities of the brain, and hence the obstructive hydrocephalus—a factor of great moment in accentuating, sometimes abruptly, the pressure symptoms which were previously inconspicuous. The fluid, which backs up in the spinal meninges, may be demonstrated under tension by a lumbar puncture, although its complete withdrawal from this source is dangerous. It may occasionally escape through the nares—cerebrospinal rhinorrhœa.

Another element, in addition to the mere size of the growth itself, must be taken into consideration, namely, the tendency of the brain itself to become œdematous—a condition which may lead to a sudden and marked increase of intracranial tension. These areas of *œdema* are often macroscopically evident on section of the brain, and are due either to vascular stasis or to local changes in osmotic pressure from chemical alterations of the cerebral fluids. They occur most frequently in the neighborhood of the growth, although they may be found at a distance, as Collier<sup>1</sup> has shown, and greatly confuse the regional diagnosis. Now these pressure disturbances, whether from swelling of the brain itself or from the encroachment of an enlarging tumor, affect the tension locally more than at a distance, although all portions of the intracranial cavity feel them to a greater or less extent. The cerebral chamber is partitioned in a measure into three subdivisions, by falx and tentorium, and a tumor in one of these compartments increases pressure locally to a greater extent than in the others, unless the tension is more or less equalized through the medium of cerebrospinal fluid obstruction. As a result of the unequal tension, distortion of the partitioning membranes, particularly of the falx, is common, with more or less dislocation of the hemispheres.

The brain, being a semifluid mass capable of certain alterations in

<sup>1</sup> *Brain*, 1904, xxvii, 490.

form, tends to escape—to herniate, in other words—through any opening which may preëxist or be made in the dense enveloping dura. As Wolbach<sup>1</sup> has shown, small, actual hernias of the cerebral substance are found over the surface of the brain, corresponding with the arachnoidal tufts—particularly along the walls of the sinuses, although by no means limited to this situation, for the cerebral surface over the temporal lobes may become dotted with these minute protrusions. There is but a single normal opening of any size in the dura; namely, at the foramen magnum, and here, particularly in case the growth originates below the tentorium, a more or less marked *foraminal hernia of the medulla* and surrounding fringe of cerebellum occurs with dislocation of these structures downward. Sudden implication of the bulbar circulation from this cause is supposed to explain the unexpected fatalities which characterize some cases of cerebellar tumor; and a lumbar puncture with the removal of the supporting column of spinal fluid is particularly apt to lead to this accident, due to the wedging of the medulla in the foraminal ring.

This tendency of the brain under pressure to herniate may sometimes by natural processes serve to relieve the general pressure symptoms, for occasionally a superficially placed tumor, especially one of dural origin, may, through pressure absorption, destroy the overlying bone and allow tumor and brain to protrude under the scalp. This is apt to occur in areas where the bone is thin, as the squamous portion of the temporals.

Changes may occur in the skull other than diastasis of the sutures and local absorption. Deepening of the Pacchionian depressions is often a marked feature accompanying the formation of Wolbach's hernias. The entire inner surface of the cranium at times becomes thinned and roughened, so that it has a sand-paper feel, and the stasis in the cranial vessels may lead to great diploëtic channels.

A word must be added concerning the respiratory and blood-pressure reactions. An intracranial growth, owing possibly to its relatively slow advancement, rarely shows the same respiratory and vascular reaction that characterizes a sudden or abrupt encroachment on the intracranial space, as in apoplexy, traumatic hemorrhage, or œdema. The slow pulse, rise in systolic pressure, and rhythmic respiration accompanying these states occur in their typical form in tumor cases only when there has been a hemorrhage into the growth or sudden œdema or hydrops ventriculorum. Otherwise one rarely sees the symptoms of acute compression in these long-drawn-out conditions except as a terminal event, for the rise in blood pressure and primary respiratory failure are the common phenomena antecedent to death.

**Headache.**—We are here confronted with a problem very similar to that in reference to abdominal pain. The brain itself, like the liver, spleen, and intestines, is insensitive, as is also its immediately investing membrane, the pia-arachnoid. The cranial cavity, however, is lined with and partitioned by an outer sentient membrane—the dura—the afferent fibres of which, trigeminal and vagal, are capable of demonstration by dissection. The fact that patients after trigeminal neurectomy—an

<sup>1</sup> *Jour. of Med. Research*, 1908, N. S., xiv, 153.

operation which produces anesthesia of the dura as well as the face—no longer suffer from the sensation of headache on the corresponding side, would favor the view that the dura plays a part at least in these subjective discomforts; but in a number of instances in which cranial operations have been performed upon conscious patients without anesthesia it has been found that the dura is actually insensitive to touch or incision and that a sensation of pain is elicited only when traction is made upon it. It would seem, therefore, that distortion of falx or tentorium and the consequent stretching of the membrane are actually the important factors.

A good illustration of headaches which may be particularly insufferable are those which occur in association with tumors or enlargements of the pituitary gland which distend its enclosing pocket of dura; and it has been found that these discomforts may cease as soon as the capsule of the gland has been incised, even though the growth be not removed. It is even better known that the headaches of tumor may oftentimes be completely relieved by a successful “decompressive” operation, and that even when a certain amount of discomfort persists after such a measure its severity varies with the degree of tension evident in the protruding area of brain. Indeed, many headaches not associated with tumor, such as those accompanying traumatic œdema, nephritis, and possibly even many cases of supposed migraine, may similarly be due to pressure.<sup>1</sup>

Headaches are not invariably present, for certain tumors need not be associated with an increase of pressure. This is notably true of many of the lesions which occur below the tentorium, such as the familiar cerebello-pontine tumor, which may give merely local symptoms possibly for years with no notable increase of tension until it reaches such a size that it obstructs the aqueduct of Sylvius, when a rather abrupt onset of pressure symptoms may occur.

When present, headache may be persistent or occur only at fairly definite hours of the day or over certain periods of time, to be followed by an interval of partial or total remission. The sensation may be nothing more than a feeling of general fulness, or it may be agonizing. Headaches are rarely definitely localized to any particular points except when a lesion immediately underlies and possibly has caused thinning of the adjacent cranium, when local pain with associated tenderness may be present. As a rule, patients refer them indefinitely to the frontal or occipital region or to the vertex more often than to one side or the other. Discomforts primarily referred to the occiput are suggestive of a sub-

<sup>1</sup> It must be admitted that many points relating to headache are not entirely explained by assigning pressure as the sole cause. Thus, lumbar puncture with the withdrawal of cerebrospinal fluid from a healthy individual—a condition that is naturally supposed to bring about a lessening of the normal tension—may occasion intracranial discomfort, and the definite reflex headaches of gastric or ocular origin are difficult to reconcile with this view. Perception of pain is unquestionably a cerebral function, and yet the brain itself (with the possible exception of the basal ganglia), so far as we know, is insensitive to the stimuli which give pain at the periphery. Thus faradization of the supposed sensory cortex in two conscious patients, although giving vague tactual impressions referred to the periphery, provoked no local sensation.—*Boston Med. and Surg. Jour.*, 1909, elxi, 71; also *Brain*, 1909, xxxii, 44.

tentorial lesion, although too much faith cannot be placed upon this, for a posterior lesion may give frontal headaches, and a frontal lesion discomfort referred to the occiput.

**Vomiting.**—Whether there is a centre in the medulla for this act—possibly vagal—can only be conjectured, but the symptom is thought to be more common in lesions involving the bulb. It is inconspicuous or entirely wanting in two-thirds of the cases, even after they have reached such an advanced stage that the evidences of pressure are pronounced. The typical so-called projectile or cerebral vomiting is common only in those cases in which headaches are severe, and is rarely seen until a late stage when pressure symptoms are pronounced; it hardly deserves the prominence given it as a cardinal symptom. When present it is often an early morning occurrence or brought on by some sudden change in position. The act may be unassociated with nausea. Unlike the other striking symptoms of pressure, it rarely occurs alone, whereas headache, choked disk, or vascular disturbances may exist almost as the sole pressure manifestation.

**Choked Disk.**—This, the most striking objective sign, is at the same time the only one of the general pressure symptoms which lends itself to experimental reproduction. Many terms have been employed in designation of the process and many views advanced to explain the causal agency: of the two most important, one has attributed the lesion to purely mechanical agencies, the other to toxic or inflammatory ones. Hence a terminology has risen which is confusing; and “optic neuritis” or “papillitis,” introduced by von Leber, as opposed to “choked disk” (Allbutt’s adaptation of the German *Stauungspapille*) and “papill-œdema,” are variously used by different writers to express the whole process or to indicate its different stages.<sup>1</sup>

Influenced by recent clinical and operative experiences, as well as by newer experimental investigations, the pendulum of opinion has swung away from the toxic toward the mechanical view. It seems in all probability a stasis œdema from the forcing of the cerebrospinal fluid into the meningeal sheath which invests the optic nerve. The sheath becomes distended, the nerve head œdematous, venous stasis occurs, and the ophthalmoscopic picture shows at this early stage tortuosity of the retinal veins with injection of the disk and more or less haziness of its outlines—the nasal margins usually being the first to become obscured.<sup>2</sup> Shieck has modified this mechanical theory in so far as to point out that the fluid leaves the meningeal sheath and passes into the optic nerve by way of the perivascular spaces which accompany the central vessels of the nerve.

<sup>1</sup> Excellent summaries in regard to the causation of choked disk have been given by Th. Kocher in *Nothnagel’s Specielle Pathologie und Therapie*, 1901, ix, 206, and by Alfred Sânger in the *Wien. med. Woch.*, 1904, liv, No. 47, pp. 2201, 2231, 2258. *Gower’s Medical Ophthalmoscopy* remains to this day the best monograph in English on this and allied subjects. Cf. Cushing and Bordley, *Jour. Am. Med. Assn.*, 1909, lii, 353, also Paton and Holmes, *Brain*, 1911, xxxiii, 389.

<sup>2</sup> This stage is usually miscalled by ophthalmologists an “optic neuritis” if it be recognized at all as pathological and sharply distinguished from “choked disk,” the term reserved for a later stage, when there is a measurable swelling of the papilla.

Marcus Gunn<sup>1</sup> divided this process, for which he suggested the unfortunate name "tumor papillitis," into the following stages:

*Stage 1.* The earliest ophthalmoscopic signs are increased redness of the disk, loss of definition in its edges, slight prominence of its surface, and narrowing of the physiological pit.

*Stage 2.* At a rate which varies much in different cases and which seems to bear a decided relation to the degree of intracranial tension, the swelling of the papilla increases, the physiological pit disappears, and the disk edges become quite obscured; along with these signs there is now slight haziness of the surrounding retina and the retinal veins show evidence of retarded circulation.

*Stage 3.* In an advancing case the next alteration consists in further swelling of the papilla, so that it becomes more prominent and occupies a larger fundus area, the venous distension becoming very marked; fine folds not infrequently appear in the œdematous retina, particularly between the disk and macula, and there may be retinal hemorrhages.

*Stage 4.* The papilla becomes more opaque and sometimes more prominent, the hemorrhages increase in size and number, and there are inflammatory exudations on the disk and surrounding retina. At this stage vision has become impaired.

*Stage 5.* The next change consists in a gradually decreasing vascularity of the papilla, parts of its surface becoming paler than normal, while the prominence either persists or slowly subsides. At this time also we first note a change in the branches of the central artery, in the form of diminished breadth—the state of atrophy with inevitable blindness.

There is every reason for ascribing all these stages to a single process; and there are reasons for regarding the process as mechanical. For a similar change occurs in the neuroretinal tissue in cases of cerebral œdema following trauma, and although this rarely advances beyond Gunn's Stage 2, still we may see the process advance even to the stage of atrophy in persisting cerebral œdemas such as those which occur in nephritis. Then, again, it is a frequent experience to see the choked disk of tumor subside after a modern palliative operation, even though the tumor be not removed, so that the toxic elements, if any exist, should still be at work.

Choked disk, when present, is doubtless one of the most valuable signs of tumor, but it must constantly be borne in mind that it is absent in all cases at an early period, in most cases until a late period, and that many tumors have a long life. The discussion as to the percentage of cases in which choked disk occurs seems therefore to be beside the mark, for, as the diagnosis is made earlier, doubtless it will be absent in an increasingly greater proportion; and we feel a certain pride in having successfully removed a tumor in a number of cases before any evidence even of congestion of the disk had set in. Tumors allowed, uninterrupted, to run their full course will all probably show a certain stage of choked disk before the end.

The advent of a choked disk may be long delayed and yet may progress

<sup>1</sup> *British Medical Journal*, 1907, ii, 1126.

rapidly when the intracranial conditions lead to its formation. It is not uncommon to see the lesion develop while the patient is under observation, and hence frequent examinations are requisite in all suspicious cases. In one of our patients, coincident with a hemorrhage into a glioma, a swelling of four or five diopters occurred within a few hours—the condition being comparable to the experimentally induced acute lesion in animals.<sup>1</sup> These acute oedemas of the neuroretinal tissues may subside after pressure has been relieved almost as rapidly as they occur; it is only in the case of a long-standing lesion that subsidence of the swelling is delayed, owing to the naturally slow absorption of the new-formed tissue which fills the physiological cup.

Choked disk may be of certain localizing value, for it is apt to make its appearance, as Horsley emphasizes, earlier on the side of the lesion. Later, or when both eyes have become involved, the mere measurable height of the swelling is not of equal moment, for the older and more advanced process may project from the level of the retina less prominently than the more recent one. Attempts have been made likewise to use choked disk as an evidence of a cerebral or cerebellar lesion, the view being taken that the subtentorial growths in a large percentage of cases have a high grade of choked disk; but this, it seems, is merely a question of the time at which the diagnosis has been made, for in the majority of the cerebellar cases the symptoms, often of long standing, are rarely thought to be due to tumor until the advent of the neuroretinal change.

One particularly important fact, properly accredited to Hughlings Jackson, is the retention of visual acuity even in an advanced stage of the process. When once vision begins to fail, owing to atrophy of the nerve from the contraction of the new tissue, the loss of acuity may take place rapidly, usually with a concentric contraction of the visual field. Vision, indeed, may be suddenly lost. It is this which speaks urgently in favor of the modern palliative operation; for if the process has advanced to Gunn's Stage 4 the prognosis for permanent vision after operation is bad, and in Stage 5 almost hopeless.

Choked disk is a sign by no means limited to intracranial tumors, for any condition which similarly and in equal degree increases pressure will naturally lead to a like neuroretinal oedema. There are certain reasons, indeed, for believing that the so-called albuminuric retinitis is merely a modified form of the same lesion,<sup>2</sup> and the same may be said of the neuroretinal change which accompanies cerebral concussion or contusion, apoplexy, thrombosis, and a number of other intracranial conditions. It is, comparatively speaking, rare in congenital hydrocephalus and meningitis.

**Choked Labyrinth.**—This condition—the *Stauungslabyrinth* of Steinbrugge—thought to bear the same relation to the auditory that choked disk bears to the optic nerves, has been much debated. It is a conjectural lesion which has not been subjected to conclusive histological demonstration or reproduced experimentally. Although many cases show a diminution of auditory acuity, there are no definite clinical means of

<sup>1</sup> Cushing and Bordley, *The Johns Hopkins Hospital Bulletin*, 1909, xx, 95.

<sup>2</sup> Byrom Bramwell, *Clinical Studies*, 1907, N. S., v, 1; Bordley and Cushing, *Am. Jour. Med. Sc.*, 1908, cxxxvi, 484.

testing for the lesion—at least nothing equivalent to the ophthalmoscope in choked disk. Batten and Collier think that stasis may affect the spinal cord and the posterior roots, especially of cervical and lumbar regions. Degenerations have been demonstrated in the entrance zone of the roots, and the condition is said to occasion pains in the root areas and loss of the tendon reflexes.

**Dyschromatopsia** (interlacing and inversion of the color fields).—Since Charcot, this has been regarded as a condition peculiar to functional or hysterical states. The perimetric examinations by Bordley and Heuer in some of the early cases of the author's series led us to believe that color interlacing was a common and early expression of an increase of intracranial tension, even before the appearance of ophthalmoscopic changes in the disks. Though this view received some support by a number of other observers, perfected methods of perimetry, particularly in regard to the character of the color peripheries, have led us to believe that our early interpretation was erroneous and that the interlaced fields were due to a combination of inattention on the part of the patient and faulty technique on the part of the observer.

**Other Pressure and General Symptoms.**—Other less important general pressure symptoms are *vertigo* and *dizziness*, possibly more common with subtentorial lesions than with those of the cerebrum and possibly also related in some way with the labyrinthine disturbances which have been described; *drowsiness*, with repeated yawning, is not infrequent when pressure is considerable and the patient in an apathetic state; *convulsions* are occasionally seen irrespective of those characterizing involvement of the precentral gyrus; and certain *psychic disturbances* with intellectual dulness are to be made out in most cases of tumor, even with no direct involvement of the frontal lobes. Rapid *loss of weight* is common, although the reverse may occur in tumors primarily or secondarily affecting the hypophysis; and *disturbances of pulse rate, respiration, body temperature, and urinary secretion* may also be regarded as general symptoms in certain cases. In some rare cases, in the absence of demonstrable vascular lesion, a loud systolic *bruit* may be heard over the cranium.

**The Focal Manifestations.**—In evidence of the gaps in our knowledge of neurological physiology, we must still refer to large areas of the brain as "silent." Fortunately for the regional diagnosis of tumors, these areas are becoming more and more constricted. The function not only of the cortex, but of deeper parts as well, is becoming sufficiently well determined to be of great diagnostic service. Irritation or paralysis, whether from simple pressure or from destruction of the various centres or paths of known function, gives symptoms which are available for this purpose, whether the lesions occur in the motor or sensory spheres, in those related to the special senses for taste, smell, vision or hearing, in the more highly differentiated area associated with the speech or stereognostic mechanism, in the psychic sphere itself, or involve the cerebral nerves in their peripheral intracranial course.<sup>1</sup> However, the matter may not be so simple as this for *symptoms at a distance*; false localizing

<sup>1</sup> For illustrative cases of tumors in the various areas Beever's *Lettsomian Lectures* may be consulted, *Trans. of the Med. Soc. Lond.*, 1907, xxx, 150 to 233.

signs which greatly confuse the primary focal and neighborhood manifestations<sup>1</sup> are apt to occur, especially late in the history of tumor.

These distant symptoms are apt to be especially confusing when the growth occupies a fairly silent area, but leads to oedema elsewhere. We have been misled by false signs in a number of instances, some illustrations of which may be given. Thus in one case a tumor of one frontal lobe so greatly indented the other as to make the latter appear to be the primary seat of trouble; a tumor of one occipital lobe in a child blind from optic atrophy, so that hemianopsia was absent, gave definite cerebellar symptoms; medullary symptoms have been far from infrequent in cases when there is a large intracranial growth situated almost anywhere; hydrocephalus has often effectually masked practically all local symptoms. A growth may become so large in a young child as to simulate essential hydrocephalus; a basilar process, particularly when luetic, may lead to vascular thrombosis with distant symptoms; nerves situated far from the lesion may be stretched and paralyzed. This is particularly common with the sixth cerebral nerve, which often becomes constricted in cases of cerebellar tumors through pressure of one of the lateral branches of the basilar artery, thereby causing diplopia.

It was made apparent in discussing the general pressure manifestations that one or another of these phenomena may possess elements of localizing value. Thus, persistent suboccipital headache and vertigo are characteristic of a cerebellar lesion; choked disk is apt to occur first upon the side of involvement in lesions of the cerebral hemispheres;<sup>2</sup> hemiachromatopsia, which may foretell a total hemianopsia, likewise may point to the hemisphere involved; and any unusual disturbances of pulse and respiration, to a lesion in the neighborhood of the medulla. These, however, are not commonly classified under focal symptoms, which concern more specifically the disturbed function of definite lobes, centres, or paths which can be conveniently taken up *seriatim* under the heading:

**Regional Diagnosis.**—This can be aided by ways other than the mere study of neurological phenomena. Thus auscultatory percussion of the shaved scalp may occasionally be helpful, the audibility of the transmitted sound of the tuning fork being lessened over the tumor, particularly if it be a solid growth or one placed near the skull. The Röntgen rays may prove of value in locating growths which have undergone calcification or have eroded or displaced the cranial bones—particularly true of tumors which deform the sella turcica—or in disclosing teratoid tumors which contain bone themselves or in demonstrating an internal hydrocephalus by the presence of pressure digitations on the bone and of separated sutures. The Neisser-Pollack method of puncture through the intact skull (*Hirnpunktion*) is not to be commended.

*Lumbar puncture* as a diagnostic measure has been much abused. On the sudden withdrawal of the supporting column of spinal fluid the

<sup>1</sup> Collier, *Brain*, 1904, xxvii, 490.

<sup>2</sup> To quote from Marcus Gunn: "Double optic neuritis with surrounding retinal change coming on quickly suggests the cerebellum; a one-sided neuritis or marked difference suggests the cerebrum, and on the whole is in favor of the tumor being on the same side as the excess of neuritis." This also is Horsley's view. Leslie Paton's studies of the same series of cases disagree with these views. (*Brain*, 1909, xxxii, 65.)



pressure from above, which continues unabated, serves to wedge the medulla and surrounding fringe of cerebellum into the cone of the foramen, with resultant anemia of the vital centres and sudden respiratory interruption. Three of the fatalities in the unoperated cases in the author's series promptly followed this measure, and the literature records many other similar experiences. The procedure in tumor cases is therefore hazardous and of no particular diagnostic value; for mere measured tension of the spinal fluid is of no significance, nor is a cytological study of the fluid or a Wassermann test of great moment in cases in which a high grade of increased intercranial tension is evident.

**Frontal Lobe.**—Lesions in this situation lead to mental disturbances, which in a way are comparable to the symptoms one meets in paresis—indifference, unpunctuality, mental enfeeblement, loss of memory and power of attention, change in disposition with more or less marked irritability or taciturnity or obstinacy or jocularity, etc., often a rambling speech, lack of realization of the illness, and change in the general conduct of life with habits of untidiness. These, in greater or less degree, characterize most of the cases, although it is often astonishing to find how inconspicuous the symptoms may be with a very extensive growth. They may often be of rather abrupt onset and not until the situation of the lesion is definitely disclosed and careful interrogation made into the patient's previous mental state is it possible to learn that in all probability some mental alteration has been of long standing.

There is undoubtedly a great difference between the two sides, the symptoms being more pronounced in left frontal lesions in the right-handed. It is to be noted that a tumor of one side may markedly affect, by indentation or compression, the opposite hemisphere. The symptoms also depend somewhat on the situation of the growth, whether it affects the prefrontal region, in which case they are said to be more pronounced, or whether it lies at the base or upon the external surface of the frontal lobe. Disturbances of the speech mechanism and of the motor activities of the contralateral side of the body may be expected when the posterior part of the lobe is affected. Vocal aphasia is common, and a good instance of the rarer agraphia has been recorded by McConnell.<sup>1</sup> There may be cortical epilepsy with movements of head and eyes to the opposite side; automatic movements are also described, and a form of frontal ataxia. Grainger Stewart<sup>2</sup> has shown that a homolateral tremor with loss of abdominal reflex on the contralateral side of the body is of frequent occurrence. Lesions here are also accompanied by relatively early nutritional disturbances.

**Temporal Lobe.**—With the exception of the uncinate area this constitutes a relatively silent region. Large tumors may give such vague symptoms that the first local sign of trouble may be the tenderness and bulging which fortells perforation and atrophy of the squamous wing. It is not uncommon in the usual subtemporal decompressive operation to expose an unsuspected temporal growth; this has occurred many times in our series. Left temporal lobe tumors in right-handed people

<sup>1</sup> *Univ. of Penn. Med. Bull.*, 1905, xviii, 156.

<sup>2</sup> *Rev. of Neurol. and Psych.*, 1906, iv, 809.

may lead to disturbances of motor speech when they encroach on the frontal area, and hemilingual or facial weakness may occur should the growth implicate the lower precentral gyrus. It is unusual for them to cause auditory disturbances, although inability to apprehend spoken language is supposed to follow a lesion of the transverse temporal gyrus of Heschl, with more or less upsetting of the entire speech mechanism.<sup>1</sup>

Cortical lesions at the tip of the temporal lobe which implicate the uncinate and possibly the hippocampal gyri are known, largely through the papers of Hughlings Jackson, to occasion certain peculiar seizures known as the *uncinate group of fits*. Purves Stewart, in 1899, was able to find reports of six typical cases, and Mills<sup>2</sup> collected as many more. The condition, must be of fairly common occurrence, for we have had many examples in our series and Kennedy<sup>3</sup> has reported a number of others.

A tumor which occasions these symptoms may either arise primarily in the temporal lobe or may originate elsewhere, as in the interpeduncular space, and involve the uncinate gyrus secondarily. The *uncinate seizures* are characterized by subjective sensations of smell or taste—the impression usually being a disagreeable one—and often by an epigastric aura. Together with these sensations, movements often occur such as those of the acts of tasting or smelling, chewing or swallowing. Salivation is not unusual. Consciousness is retained, as a rule, in these attacks, and they are not infrequently replaced by or associated with so-called “dreamy states,” described as a distressing sense of unreality of surroundings. Respiratory phenomena have been noted. Neighborhood symptoms occur. In one of our patients there were secondary hypophyseal disturbances; in another, a fluctuating contralateral hemianesthesia and homonymous hemianopsia due to pressure against the adjoining crux and visual pathway were present—a combination of symptoms which had led to a diagnosis of hysteria, for the tumor proved to be a benign one and had been present for many years. We have found in temporal lobe lesions that disturbances of the visual fields are common, sometimes as an upper quadrantal homonymous defect. Hemianopic visual hallucinations may occur.<sup>4</sup>

**Paracentral Convulsions.**—For evident reasons by far the greater number of tumors which have been localized in the past have involved the central motor and sensory gyri. A small growth which implicates the surface of this part of the brain almost invariably leads to irritative symptoms, beginning either with a primary motor or sensory aura in leg, body, arm, neck, face, or tongue, as the case may be. These focal or Jacksonian seizures may for a long time be the only manifestations; they may remain limited to the part involved or may spread with a typical “march,” so as to involve the entire body in convulsive move-

<sup>1</sup> The rarity with which word-deafness occurs in cases of involvement of the superior temporal gyrus by tumor or by the protrusion of the lobe through a palliative defect makes the cortical representation of this primary word centre a matter which would seem to deserve as careful consideration and possible revision as has been given to Broca's vocal speech centre since Marie's recent criticism; cf. Adolf Meyer, *Jour. of Psychol. and Neurol.*, 1908, xiii, 203.

<sup>2</sup> *Jour. Am. Med. Assn.*, 1908, li, 879.

<sup>3</sup> *Arch. Int. Med.*, 1910, viii, 317-350.

<sup>4</sup> A. Pick, *Am. Jour. Med. Sc.*, January, 1904.

ments with loss of consciousness. As the lesion enlarges it is characterized by irritative disturbances to be followed by paralytic ones, shown first in the musculature of the part in which the attacks originally began. The more superficial the lesion the more circumscribed may be these paralyses; the deeper the lesion and the nearer the internal capsule the more extensive they are.

It must be borne in mind that the cortex of the central fissure dips down to a depth possibly of an inch; also that, as Sherrington and Grünbaum's observations have shown, the motor strip extends to the depth of the fissure, so that a lesion truly cortical may nevertheless lie at a considerable depth below the surface of the hemisphere. Sensory anesthesia is much less likely to follow primary sensory fits than in motor palsy to follow either sensory or motor seizures; for although the cortical terminals for common sensation have with some definiteness been placed in the gyrus centralis posterior, still there are several relays between periphery and central end-stations, and a fairly extensive cortical or subcortical lesion is necessary before definite areas of anesthesia become demonstrable.

The most characteristic symptom of tumor in this situation—if one wishes to delay intervention until it occurs—is the gradual transference of a local convulsive movement into a local paralysis of movement. The paralytic features occur first as a mere exhaustion after the attack in the affected muscular territory and then as an actual paralysis coincident with destruction of the centre. After this, convulsions no longer occur in these muscles, but in those whose centres are adjoining. The first convulsion is usually local, rarely a general one, although soon local and general convulsions become interchangeable. Not infrequently between the attacks continuous twitching—"epilepsia continua"—may occur. It is the usual rule for all objective sensory disturbances to be wanting in the paralyzed area, although it is not rare to find certain disturbances of tactual sense or of the sense of localization.

**The Parietal Lobe.**—A lesion of the superior lobule, particularly if subcortical, leads to a contralateral disturbance of the stereognostic sense.<sup>1</sup> If the subcortical lesion be large and encroach on the postcentral gyrus, it may occasion additional sensory disturbances, such as loss of muscle sense, of position in space, or even of some forms of common sensation. Consequent upon the sensory disturbances certain irregularities of moment appear (the motor apraxia of Leipmann). The arm, for example, although not in a strict sense paralyzed, for the patient when observing the member can make powerful though somewhat awkward movements, yet cannot be used for any purposeful act, particularly if the patient be blindfolded. More or less muscular rigidity accompanies the condition, although it is unassociated with any direct implication of the pyramidal tract.

One characteristic symptom elicited by a tumor involving the left angular gyrus in the right-handed is the inability to appreciate written language or even to read letters—word-blindness (*alexia*). A deep-

<sup>1</sup> Although much discussed, astereognosis seems to be a fairly definite and reliable symptom of lesions of the superior parietal lobule. A most typical instance in a case of stab wound—not tumor—has been recorded by the writer in the *New York Med. Jour.*, 1907, lxxxv, 161.

seated lesion of either lobule may involve the fibres of the optic radiation and lead to a half-blindness of the corresponding halves of both retinas, either for colors alone or for form also—homonymous *hemiachromatopsia* or *hemianopsia*.

The right parietal lobe represents a comparatively silent area, formerly regarded by Sanger as the most favorable for decompressive operations.

**The Occipital Lobe.**—A crossed homonymous hemianopsia is the characteristic, and may persist for a long time as the only, symptom in addition to those incidental to general pressure. If optic atrophy secondary to a choked disk has occurred, and there is no history of half-blindness before the loss of vision, a regional diagnosis may be impossible. Occasionally the fields for color may be affected before those for form suffer any restriction, and cases have been recorded in which quadrantal blindness has been present, although these fractional defects in the field are possibly more characteristic of implication of the visual pathway farther forward as it traverses the temporal lobe.

Homonymous hemianopsia, of course, is characteristic of a unilateral lesion of the visual pathway in any part of its course, from occipital lobe to chiasm, and neighborhood symptoms must be depended upon to determine the seat of the lesion. When the occipital cortex, particularly the more important region of the calcarine fissure, is involved, the loss of half-vision may be ushered in by certain visual hallucinations, such as scintillating scotoma. Involvement of the optic radiation itself on the left side may be accompanied by some of the parietal lobe symptoms just enumerated—alexia or optic aphasia. Lesions farther forward, which include the fibres from the oculomotor nuclei on one side and affect the arc of the pupillary light reflex (Plate V), lead to the hemianopic loss of this reflex on the blind side (Wernicke)—a condition which is wanting in pure occipital lesions. Occipital lobe tumors which have reached a great size may produce pontine disturbances or even symptoms indicating some implication of the corresponding cerebellar hemisphere.

**Centrum Semiovale and Basal Ganglia.**—Needless to say, lesions in any of the situations heretofore considered involve to a greater or less extent the subcortical areas, and it is often difficult to determine from the symptomatology alone whether a tumor is entirely central or whether it actually reaches the cortex. This is the more so since the cortical areas represented by the insula and by the deeper fissures dip down well into the central parts of the hemispheres, and some cortical manifestations may be present in many of these lesions, even though they lie far beneath the actual exposed surface of the brain. It is chiefly by the relatively large extent of the contralateral paralyses that one is enabled to conjecture the approximate depth of the lesion. Particularly in the left half of the brain the deeper lesions of the centrum lead to very extensive disturbances not only in movement and sensation but in psychic activity, in the speech mechanism, and in vision.

Tumors of the *basal ganglia*, particularly those of the thalamus, are often capable of localization, the so-called *thalamic syndrome* having received much attention of late. The most characteristic symptom of a thalamic lesion is a contralateral disturbance of movement, either in the

# PLATE V

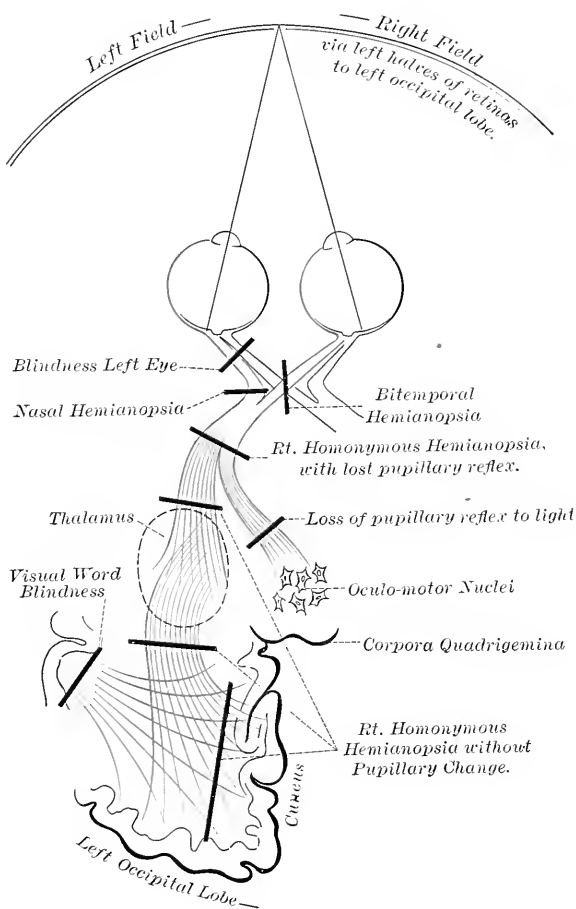


Diagram to Show various Forms of Visual Disturbance following Lesions in Different Portions of the Left Visual Pathway.



nature of a coarse tremor or of choreic or athetoid movements, often accompanied by painful subjective paresthesias. Indeed, this is the only area of the brain a lesion of which seems to produce subjective sensations of discomfort referred to the periphery. Roussy<sup>1</sup> outlined this syndrome as follows: The patients present (a) persistent hemianesthesia, more or less marked in modalities of touch, pain, and temperature, but always pronounced in regard to deep sensation; (b) slight hemiplegia of a transient nature without contractures; (c) slight hemiataxia and more or less complete astereognosis; (d) persistent paroxysmal pains on the affected side, often intolerable and unrelieved by any analgesic remedy (*hémiplegie douloureuse*); and (e) athetochoreic movements on the affected side.

In addition to these, the most characteristic phenomena, neighborhood symptoms occur, such as contralateral hemiplegia; oculomotor palsies, such as crossed pupillary dilatation and ptosis; emotional palsy of the face; hemianopsia; deafness from involvement of the lateral geniculate bodies, and so on.

In the hemiplegias which are due to lesions of the cortex or subcortex exaggeration of the deep reflexes and a persistent Babinski sign, together with contractions and epilepsy, are to be expected, with an absence of the athetochoreic movements and subjective painful sensations.

**Corpus Callosum.**—Bruns found only 26 recorded cases of tumor primarily involving this structure, and it is unusual for them to give symptoms sufficiently definite to insure a local diagnosis, although this has been correctly made in a few instances.<sup>2</sup> In addition to the usual pressure symptoms, these lesions are accompanied by a slowly progressive, bilateral, though not necessarily symmetrical, palsy of both extremities, without paralysis of the cerebral nerves, the latter being regarded by Raymond and others as an important negative symptom. Particularly when the anterior portion of the corpus callosum is involved, intellectual disturbances are marked,<sup>3</sup> often with apraxia and alterations of the speech mechanism. There is apt to be a profound stupor, and the palsy of the extremities affects movement of the legs more than of the arms. Sensory disturbances are absent and the reflexes normal. The sphincters are uncontrolled. Tumors which involve the median portion of the corpus lead to more marked disturbances of motion, and a form of callosal ataxia has been described. Tumors which involve the splenium are apt to implicate adjoining structures with neighborhood symptoms.

**The Brain Stem (Crura, Corpora Quadrigemina, Pineal Gland, Tegmentum and Pons.)**—It is only natural that distant and neighborhood symptoms, as well as complex local ones, occur with lesions in this situation—the “cross-roads” of the nervous system. Thus pressure upon or involvement of the internal capsule or cerebral peduncles produces a more or less marked contralateral hemiplegia, possibly with some degree of hemianesthesia; and if the growth extends outward from the mid-brain hemi-

<sup>1</sup> La couche optique (étude anatomique, physiologique et clinique); le syndrome thalamique. Steinheil, Paris, 1907, p. 371.

<sup>2</sup> Putnam and Williams, *Jour. of Nerv. and Ment. Dis.*, 1901, xxviii, 645.

<sup>3</sup> More than with tumors of the frontal lobes themselves, according to Knapp, *Brain*, 1906, xxix, 35.

anopsia may occur, and likewise the muscular sense and stereognosis may be affected. The deep reflexes may thus be increased with absence of the superficial ones—this being particularly true of the Babinski phenomenon. Any growth in this situation will in the course of time lead to an internal hydrocephalus, which greatly complicates the symptom complex, in consequence of the widely distributed pressure against the hemispheres.

Several examples of tumors, often a solitary tubercle, of the *crura* have been recorded<sup>1</sup> the characteristic symptoms being a contralateral hemiplegia, with homolateral or bilateral oculomotor palsies.

Involvement of the *corpora quadrigemina* leads to a staggering gait with unsteadiness and deviation sometimes to one side or the other in progression, and an especial tendency to fall directly backward; also to a failure of sight and hearing when, as the growth spreads to the side, the lateral geniculate bodies become invaded or compressed; to nystagmus; to persisting palsies of the ocular movements, particularly those in a vertical direction,<sup>2</sup> with preservation of lateral movements; and possibly to loss of the pupillary reflex to light and even to convergence as the centres for the oculomotor nerves become involved. The pupils may be irregular; and Bielschowsky describes a form of nystagmus with clonic twitching. Thus a combination of ocular paralyses and ataxia (Nothnagel) especially characterizes lesions in this situation, whether they arise in the *pineal gland* or *corpora quadrigemina*.

*Pineal* tumors occur chiefly in the young and are characterized by internal secretory disturbances which manifest themselves by a precocious adolescence, the secondary characters of sex often being acquired at an early age—at two years in one of the author's cases. At the same time adiposity and skeletal overgrowth often occur. In addition the usual symptoms of a tumor of the neighborhood appear, oculomotor palsies, deafness, ataxia and so on, together with an internal hydrocephalus from occlusion of the *iter*.

When the *tegmentum* is implicated, particularly the nucleus ruber and the superior cerebellar peduncle leading to it (the *cerebello-rubral system*), a particular syndrome is apt to be called out in addition to the above, characterized by a peculiar coarse, oscillatory tremor, described by Gordon Holmes.<sup>3</sup> If the red nucleus of one side is involved the tremor is contralateral. We have had one or two striking examples of the condition. In addition to this tremor, which Holmes likens to that of paralysis agitans, a mask-like appearance of the face occurs; and the oculomotor palsies above mentioned, with staggering gait, etc., commonly occur. The disorders of movement with posterior thalamic lesions are more apt to be of an athetoid character than in tegmental lesions.

The *pons* is not an uncommon seat, particularly of tubercles and gliomata. The latter may continue to be traversed by intact fibres,

<sup>1</sup> J. H. W. Rhein, *Jour. Am. Med. Assn.*, 1914, lxiii, 1662.

<sup>2</sup> "Persisting paralysis of associated lateral movement indicates a lesion of the posterior longitudinal bundle; of associated vertical movement, a lesion in the vicinity of the oculomotor nucleus; paralysis of associated movement does not result from lesions of extracerebral fibers." Spiller, *Paralysis of Associated Movements of the Eyeballs* (Blicklähmung), *Jour. of Nerv. and Ment. Dis.*, xxxii, p. 417.

<sup>3</sup> *Brain*, 1904, xxvii, 327.



so that they may reach an astonishing size without producing such symptoms as would be expected of a lesion here. The characteristic feature, however, of most pontine tumors is the well-known alternating or crossed hemiplegia, with nuclear cerebral palsies on the side of the lesion due to direct implication of the centres, and a contralateral palsy of the extremities with the usual evidences of pyramidal tract degeneration.

Various forms are recognized, possibly the most common combining a homolateral facial palsy with a crossed involvement of tongue and extremities. The facial territory may be the first to be affected, the brow often escaping; or, on the other hand, the palsy of the tongue and extremities may be primary, depending on the seat of the lesion. In another type the trigeminus may be affected on the side of the lesion with anesthesia of one side of the face, and there may be a coincident crossed anesthesia, pure or dissociated, affecting the paralyzed limbs. Another form may show itself, possibly in the absence of facial or hypoglossal disturbances, as an abducens palsy, crossed with a motor involvement of the limbs; and in these cases there is a characteristic paralysis of conjoint ocular movement toward the side of the lesion. Abducens palsies must be scrutinized carefully, however, as they are the commonest of all cerebral nerve lesions, and may, from pressure or from stretching through dislocation, accompany a tumor in almost any situation.

When a pontine growth involves the main conducting paths or the nuclei on each side, these typical crossed paralyses will be wanting, and some evidence of bilaterality of the lesion may in many cases be expected before the end. We consequently may have a multiplicity of combinations with unilateral or bilateral disturbances of facial, trigeminus or abducens, and it is even possible to have a coincident cerebral paralysis of all four extremities and both hypoglossi. The variations are too many to enumerate, although Bruns brings them into four main categories: (1) Bilateral paralysis of the cerebral nerves and extremities; (2) bilateral cerebral nerve with unilateral paralysis of the extremities; (3) unilateral cerebral nerve and bilateral paralysis of extremities; (4) bilateral cerebral nerve paralysis without involvement of the extremities.

Neighborhood symptoms are shown, if the growth presses forward, by oculomotor symptoms; if backward, by affections of the medullar nuclei, evidenced by disturbances of deglutition or phonation—the bulbar palsies. Salivation, albuminuria, polyuria, and extreme degrees of pyrexia have been described in association with pontine tumors. In pontine lesions a choked disk is notably late in its appearance or possibly may seem to be so owing to the fact that the diagnosis of a lesion in this situation is apt to be made early.

**The Medulla Oblongata.**—The complexity of paths and centres crowded here into a small space naturally makes the symptom complex most variable; nor are the lesions sufficiently common to enable us to more than sketch their clinical picture. The centres for the cerebral nerves, from the eighth to the twelfth, may be affected so that deafness or Ménière's syndrome, palsies of the palate and pharynx with dysphagia, vocal cord palsies, disturbances of cardiac or respiratory activity, dysarthria from weakness or paralysis of the tongue, and trigeminal anesthetics (ascending

root) may occur in combination with motor and sensory, crossed or uncrossed disturbances of the extremities. The medulla may be affected as a distant symptom of most intracranial tumors, owing to the crowding of the medulla into the foramen.

Tumors of the *fourth ventricle* occasionally give symptoms sufficiently characteristic to justify a focal diagnosis. In the greater number of cases observed the lesion has been due to a cysticercus, less often to a glioma, this being the usual seat of the ependymal glioma already described. Stern<sup>1</sup> collected 72 cases of the former and about 50 cases of tumor. The symptoms are suboccipital pain, a peculiar bowed position of the head, a periodic occurrence of general pressure symptoms, often of abrupt onset, accompanied by severe headache, vomiting, and cervical rigidity. In some patients a changed position of the head may cause a sudden accession of symptoms (Bruns); this is supposed to indicate an unattached and movable cysticercus. An interval of complete freedom from symptoms may follow. Sudden death is apt to occur during some one of the periods, with a slowed pulse and respiratory failure. Choked disk is rare. Polyuria, polydipsia, or glycosuria may be present. The fluctuating intensity of the symptoms is presumably due to a varying degree of obstructive hydrocephalus.

**Cerebellum.**—Inasmuch as the symptoms which they elicit bear a close relationship, all tumors situated beneath the tentorium which affect one or the other cerebellar hemispheres or the vermiform process, either through pressure or by direct involvement, will be considered together. According to the classification of Stewart and Holmes, in their paper<sup>2</sup> based on a series of 40 cases observed at the National Hospital, of which number the diagnosis was confirmed in 22, these growths are considered under the headings (1) of *extracerebellar*, and (2) of *intracerebellar* tumors.

Tumors here, as in other parts of the brain, lead first or last to the cardinal symptoms of pressure—headache, vomiting, and choked disk—and very frequently to vertigo or dizziness. The headache is apt to be suboccipital, often on the side of the lesion, and not infrequently is associated with tenderness on pressure. The choked disk is supposed to be found in a much higher percentage than is the case with growths elsewhere, probably due to the fact that in the course of time a cerebellar tumor will almost certainly lead to an obstructive hydrocephalus, in consequence of which a neuroretinal oedema is inevitable and may occur abruptly, often with a rapid augmentation of general pressure symptoms. In the absence of this complication, and with a slowly advancing benign lesion, choked disk may be absent for years. Vertigo is more frequently associated with subtentorial than with supratentorial tumors. When marked, the patient experiences a sensation of the movement of self or objects. According to Stewart and Holmes, in both extracerebellar and intracerebellar tumors the patient often has the impression that objects are moving in a direction away from the side of the lesion: whereas he feels himself revolving, in the case of an intracerebellar tumor, in the same

<sup>1</sup> *Deut. Zeit. f. klin. Med.*, 1907, lxi, 64; *Deut. Zeit. f. Nervenheilk.*, 1908, xxxiv, 195.

<sup>2</sup> *Brain*, 1904, xxvii, 522.

direction with the objects; in the case of an extracerebellar tumor he appears to be turning in the opposite direction.

A tumor arising in the substance of the cerebellum (*intracerebellar*) leads to symptoms which affect the musculature on the side of the body homolateral to the lesion, and occasionally to the peculiar convulsive seizures described by Hughlings Jackson and known as "cerebellar fits."

A tumor arising from the meninges and compressing the cerebellum secondarily (*extracerebellar*), of which the most characteristic type is the meningeal endothelioma originating in the lateral cerebellopontine recess, leads to more or less pressure disturbance of the cerebral nerves on the side of the lesion, in addition to the symptoms of an intracerebellar growth.

A large number of these extracerebellar tumors are supposed to grow from the flocculus or from the sheath of the auditory nerve, and as they enlarge they destroy and press to the side the pons and markedly indent the anterior and lateral portion of the adjoining cerebellar lobe. Consequently the acousticus is the nerve most commonly affected, and either subjective noises or complete nerve deafness may occur. The x-ray may show an enlargement of the internal auditory meatus. In view of its close proximity to the eighth nerve the facial may likewise be affected with weakness of its musculature on the side of the lesion. Even in the presence of a large tumor, however, it may be found stretched out to double its normal intracranial length, with little more evidences of weakness than is shown by a less complete closure of the eye in winking. A particular triad of symptoms has been pointed out by Oppenheim, namely, disturbance of hearing, paralysis of winking, and diminished corneal reflex. The trigeminus is occasionally affected, more especially its sensory portion, with pain or tingling or dysesthesia over its particular skin field. The abducens may likewise suffer and more rarely the glossopharyngeal, vagus, or hypoglossus. In intracerebellar tumors it is, relatively speaking, unusual for these nerves to be affected.

Disturbances of the body musculature are common in both intracerebellar and extracerebellar forms. Tremor, loss of muscular tone, even paresis of the limb and trunk muscles, weakness of the conjugate movements of the eyes, nystagmus, and staggering gait with tendency to fall, oftentimes toward the side of the lesion, and alteration of the deep reflexes, all occur. These symptoms are most pronounced on the side of the lesion. The ataxia, which may be very marked, is a coarse movement accentuated when the patient attempts to perform some delicate act; unlike the ataxia of tabes it does not increase when the eyes are closed—an evidence of its central origin. There is an associated condition, which Babinski termed *adiadokokinesia*,<sup>1</sup> in which the patient cannot repeat rapidly executed movements, such as pronation and supination of the forearm, owing to the defective coördination. Loss of tone in the muscles (*asynergia*) may be present, so that the arm and the leg may be completely flaccid with retention of the deep reflexes.

The ocular movements are also affected in ways characteristic of

<sup>1</sup> *Rev. neurologique*, 1902, x, 1013.

cerebellar disease. There is apt to be difficulty in forcing and retaining conjugate movements of the eyes toward the side of the lesion, and the attempt usually elicits nystagmoid movements. It is not infrequent for *nystagmus* to be present on looking toward as well as away from the presumed site of the lesion, but as Stewart and Holmes pointed out, the jerking movements are apt to be more rapid on one side than the other, the slower rhythm being usually upon the side of the lesion. Nystagmus may nevertheless be wanting. It was absent in 11 out of 44 consecutive certified tumor cases in our series.

The position of the head is apt to be toward one side, usually facing away from the lesion, and tilted back so as to relieve the strain on the cervical muscles. Alfred Gordon asserts that forcible movement of the head toward the lesion increases the headache and vertigo in a characteristic way. Irregular *tremors* are often present in the extremities. They are accentuated with the patient erect, his arms and hands held horizontally extended and in a forced position. A staggering gait is common, with a tendency to deviate, totter, or actually fall toward one side—more often toward the side of the lesion. A method of determining the affected side is to have the patient stand on one leg or the other, the chief disability appearing on the side of the lesion. Unsteadiness is equally characteristic of a tumor involving the middle lobe or vermis and one involving the hemispheres, although with the former there is a greater tendency to fall directly backward or forward.

The reflexes are apt to be affected, disparity between the two sides being of chief importance, for there is considerable difference of opinion as to whether they are increased or are diminished on the side of the lesion. It is not uncommon for them to be diminished at one time, and at a later period increased, depending in all probability upon the degree of advancement of the process. The superficial reflexes are not greatly altered, although occasionally, in association with an increase of the deep reflexes, an extensor toe phenomenon may occur.

Though the localizing diagnosis of a tumor in the cerebellum is possibly the easiest of all, nevertheless there are cases, even when a large tumor is present, in which few if any of the characteristic signs and symptoms can be elicited.

**The Cranial Base.**—A variety of tumors, either of meningeal or cranial origin, may arise from one or another of the cranial fossæ and produce definite focal symptoms, owing chiefly to the involvement of the cerebral nerves, in addition to the evidences of general pressure. Syphilitic processes are common here. Bony or cartilaginous tumors originating in the accessory nasal sinuses may invade the cranial chamber. Epithelial tumors may grow inward from the deeper ear; such a case occurred in our series, with the unilateral involvement of all the cerebral nerves from the sixth to the twelfth inclusive. The *cavum Meckelii* is not an uncommon seat for tumors which may give all the pain of a severe trigeminal neuralgia during the process of stretching or destruction of the Gasserian ganglion; and keratitis is a frequent accompaniment of the process. But of all the basal and extracranial lesions, those of chief interest and moment arise in the neighborhood of and implicate the—

**Hypophysis Cerebri.**—This is a favorite seat of tumors, both congenital and acquired. Two separate factors must be considered with tumors in the interpeduncular region, commonly grouped as hypophyseal tumors. First, what is their effect, if any, on the gland itself; and second, what are the chief neighborhood symptoms? The former has heretofore received scant attention, but the neighborhood symptoms are well recognized and have the following characteristics: Any mesially placed tumor in the interpeduncular space naturally presses upon the optic tracts or chiasm, leading in the majority of cases to partial amblyopia and to primary optic atrophy. This in most striking form occurs as a bitemporal hemianopsia, although by no means is this the only form of perimetric defect, for an homonymous hemianopsia may occur, or one nerve may suffer much more than the other; so that, for example, total blindness in one eye may be associated with normal vision or some stage of hemianopsia in the other.

Choked disk is usually absent, as there commonly is no great increase in intracranial tension, although it may occur later should the growth enlarge so as to push its way into the third ventricle and obstruct the foramina of Munro. Headache is often a common and distressing symptom, due rather to the distension of the dural pocket of the gland than to any general increase of the tension. The x-rays are particularly helpful in the diagnosis of these cases, for in many of them there is a characteristic deformation of the sella turcica; or the growth itself, if a teratoma, may cast a shadow.

The glandular symptoms are quite another matter. Since Marie's discovery of the frequent association of hypophyseal enlargements with acromegaly there have been innumerable reports of cases in which a tumor of the gland or its neighborhood has been unaccompanied by evidence of "Marie's disease." These findings have usually been interpreted as contradictory of his views. An especial group of these cases, as shown by Fröhlich,<sup>1</sup> is characterized by a peculiar tendency to the deposition of fat with sexual infantilism, hypotrichosis, etc.

The hypophysis has been proved by Paulesco to be a gland essential to the maintenance of life, its total removal leading to fatality, with a peculiar symptom complex—*cachexia hypophyseopriva*. In reading Paulesco's experiments Crowe, Homans, and the writer observed that partial removal of the gland in adult animals led to adiposity and to atrophy of the ovaries or testes,<sup>2</sup> whereas in puppies skeletal and sexual infantilism persisted. Thus the syndrome of Fröhlich was shown to be due to a condition of hyposecretion (*hypopituitarism*). On the other hand, acromegaly, as shown by the improvement in the condition after hypophyseal operations with partial removal of the gland (Hochenegg; Cushing), would appear to be due to an oversecretion (*hyperpituitarism*).

Both of these conditions may be associated with tumor, although it is probable that in varying degrees of hyper- or hypo-activity they are of common occurrence—as common as the over- or underactivity of any

<sup>1</sup> *Wien. klin. Rundschau*, 1901, Nos. 47, 48.

<sup>2</sup> H. Cushing, *Jour. of the Am. Med. Assn.*, 1909, liii, 249.

ductless gland, the thyroid for example. In the case of overgrowth (gigantism when starting early in life; acromegaly when in adult age), hypertrophy or adenoma of the gland itself is usually found. In the cases characterized by "adiposogenital degeneration" (showing persistence of sexual infantilism when originating before adolescence; and a tendency to revert to this condition when beginning in adult life) the tumor may either implicate the gland directly or may be situated above it in the infundibular region and lead to a pressure atrophy. There are other symptoms caused by these interpeduncular tumors, such as polyuria, glycosuria, amenorrhœa, impotence, psychic disturbances and so on, which in all likelihood are bound up with the activities of the other ductless glands. It is not unlikely, in view of Herring's discovery, that the hyaline secretion of the posterior lobe normally discharges into the cerebrospinal fluid of the third ventricle, that disturbances of hypophyseal activity may occur first or last in many cases of intracranial tumor. To this may be attributed the adiposity so often accompanying cerebellar tumors, particularly in children.

**Differential Diagnosis.**—The three main questions which must be answered by the clinician when confronted with a suspected case of intracranial tumor in the order of their relative importance are: (1) Is a growth actually present? (2) If so, what is its presumable situation? (3) What is its nature? It cannot be emphasized too strongly that a brain tumor is rarely diagnosed in general practice until the onset of the major symptoms of pressure—the familiar triad of headache, vomiting, and choked disk. The ophthalmologist is too often the first to suspect a tumor. It is important to appreciate the fact that tumors in the retrospect may prove to have been of years' duration before the onset of this unmistakable general pressure syndrome—an indication of the attention which must be paid in the future to the premonitory symptoms.

The early mistakes in diagnosis make a most disconcerting professional record. In our early series the majority of the cases gave a history of having undergone treatment for long periods for an astonishing variety of presumed maladies, and, it may be added, inevitably for syphilis. The usual mistakes are naturally attributable to an effort to ascribe the more or less vague disturbances antecedent to the general pressure symptoms to some cause other than the unsuspected intracranial lesion. Diagnoses of hysteria, of psychoneurosis or neurasthenia, or of gastric or ocular reflex headaches are particularly common.

**Hysteria.**—Certain of the so-called functional disturbances are inevitably superimposed on every organic lesion, and when the primary seat is obscure, and particularly if it affects the nervous system, the functional superstructure may so overtop the basal trouble as to effectually conceal it. One of our patients—the wife of a physician—for twenty years had suffered from irregular headaches, periods of amenorrhœa, intermittent grades of hemianesthesia and hemianopsia, inversion of the color fields and peculiar dreamy periods of semiconsciousness. Although seen by many during this period there was but one diagnosis—hysteria—until a terminal choked disk betrayed the nature of the lesion—a benign tumor of the right temporal lobe and uncinate region.

In an excellent paper on brain tumor diagnosis Russell<sup>1</sup> says that "the signs of organic disease cannot be confounded with anything that hysteria is able to furnish." This is true of a late period in the disease, not of an early one. The presence of organic changes, shown by a choked disk, cerebral nerve palsies, loss of the pupillary reflex, absence of the knee-jerks, an extensor plantar response, typical Jacksonian fits, and the like, serves to eliminate hysteria; but in the absence of these definite symptoms and in the presence of the so-called functional ones, brain tumor is often most difficult to rule out. Hysterical hemiplegia may at times be distinguished from organic disease by Hoover's test, which consists of an involuntary downward pressure of the supposedly paralyzed limb when the patient attempts to raise the sound member against resistance.

**General Paralysis of the Insane.**—This is especially liable to be confounded with frontal lobe tumors, particularly when the optic nerves have become atrophied, although a choked disk is said never to occur. The symptoms of tumor are usually progressive, those of general paralysis fluctuating; and the Argyll-Robertson pupil and characteristic defects of articulation are usually conclusive.

**Disseminate Sclerosis.**—This may simulate tumors of the cerebellum and mesencephalon. There may be headache and choked disk; optic atrophy is common. The vertigo, vomiting, cerebral nerve palsies, nystagmus, incoördination and spastic condition of the extremities as well as the dysarthria may occur in both.

**Vascular Lesions.**—These are often most difficult to distinguish. Pressure symptoms may be present, and local signs are equally common. In arteriosclerosis the retina often proves a tell-tale should vascular disease, in the absence of choked disk, be marked in the retinal arteries. When a combination of tumor and cerebral arteriosclerosis is present a diagnosis may be very difficult. In our series there was a patient aged seventy-two, with transient aphasia and focal seizures involving the face, in whom existent vascular disease could well have accounted for the symptoms, but an exploration revealed a small primary glioma of the lower precentral region.

**Cerebral Thrombosis.**—This may be difficult to distinguish from tumor when the process is slow, or, on the other hand, when tumor symptoms happen to be of fairly rapid onset. The vascular lesion leads to areas of softening which become œdematous and thus increase tension, so that headaches and choked disk, with the local manifestations, make the intracranial symptoms indistinguishable. This is especially true of the fairly acute forms of syphilitic encephalitis with thrombosis.

**Apoplexy.**—Here, also, the characteristic sudden onset of symptoms speaks against tumor, although it must always be borne in mind that the hemorrhage may have occurred in the substance of a vascular growth. "Stroke" in a young individual with no evidence of general vascular disease and particularly when there has been a history of headaches, must arouse a suspicion of tumor.

<sup>1</sup> *Brit. Med. Jour.*, 1907, ii, 1120.

**Aneurisms.**—Aneurisms of the larger vessels, particularly of the internal carotid, basilar, and middle cerebral, are not exceptionally rare, and may reach a considerable size without symptoms; on the other hand, they may closely simulate tumor. This is particularly true of carotid aneurisms which are found in the interpeduncular space near the chiasm, which may lead to hemianopsia, optic atrophy, or oculomotor palsy, in addition to the severe headaches simulating those of pituitary tumors. They occasionally may be recognized by a murmur, but this is less characteristic of the simple saccular aneurism than of the easily recognized arteriovenous variety. They may occur at any age, even in childhood, although naturally they are more common in adult life. Usually small, they may, as in Bramwell's case, reach the size of an orange. Their blood-content tends to clot, and may organize with spontaneous healing. Gowers gives the following series in order of their frequency: Sylvian, basilar, internal carotid, artery of the corpus callosum, posterior and anterior communicating, vertebral, posterior cerebral, inferior cerebellar artery.

When headache is present it is apt to be severe, and is increased by straining. Convulsions and vomiting may appear, but choked disk is rare, and a bruit is exceptionally uncommon except in the arteriovenous form, which could hardly be mistaken for tumor. The only patient in our series in whom a subjective and objective cranial bruit was present proved to have a contralateral glioma, and there was no aneurism. A bruit should be regarded as aneurismal only after the most careful consideration, for it may be produced by a vascular tumor, whether from its own circulating blood or by pressure against one of the large basal vessels. They are rarely diagnosed during life, and usually terminate in rupture with sudden death. In only 37 per cent. of the 555 cases studied by Beadles<sup>1</sup> did aneurism give symptoms during life, and in 67 per cent. death was preceded by an apoplectic seizure.

**Nephritis.**—The symptoms of intracranial œdema secondary to renal disease may be indistinguishable from tumor. In both, headaches, vomiting, and neuroretinal changes may be present, and focal paralyses or convulsions may accompany the nephritic œdemas—for the process is apt to be more or less restricted and need not be general in character. The so-called "albuminuric neuroretinitis" is regarded by Bramwell,<sup>2</sup> Bordley, and the writer as a process due largely to the same factor which produces choked disk, namely, pressure. We have seen the characteristic stellate figures due to exudates in many cases of tumor; on the other hand, they may be completely wanting in nephritis. *Uremia* may have many features in common with the late stages of a new-growth.

**Abscess.**—Wanting a clear history of an infected cranial wound, of otitis media or infection of the accessory nasal sinuses, a differential diagnosis may be difficult in the extreme. On three occasions we have mistaken abscess for tumor. Chronic abscesses have thick walls, and when exposed may be easily shelled out from the brain. They may be of long duration, with no antecedent history, and give symptoms the exact counterpart of a new-growth. Even a high grade of choked disk, which is

<sup>1</sup> *Brain*, 1907, xxx, 285.

<sup>2</sup> Byrom Bramwell, *Clin. Studies*, N. S., 1905, iii, 183.



thought to be rare, may accompany them. In the retrospect, however, it becomes apparent that the symptoms are more apt to be fluctuating than is the case with tumor. Leukocytosis and fever may be absent, even though viable organisms can be recovered after some months from the cavity of the more or less dormant abscesses.

**Serous Meningitis or Ependymitis.**—Serous meningitis or ependymitis leading to hydrocephalus may give a definite tumor symptom complex, although after the rather acute onset the symptoms are more apt to be fluctuating in severity, often showing long periods of abeyance. In one of our cases an obstruction of the left foramen of Munro occurred from ependymal inflammation, with symptoms indistinguishable from a tumor of the left hemisphere. Doubtless many of these peculiar cases are capable of spontaneous recovery, and should be included in the group of false or pseudo-tumors. It may be particularly difficult to distinguish a subtentorial tumor from circumscribed meningitis serosa. In addition to the pressure symptoms the latter may show nystagmus, paresis of an external rectus muscle, usually the right (Oppenheim), an inactive corneal reflex, tinnitus, and diminished hearing and cerebellar ataxia.

It is in this latter group of cases that a differential diagnosis between cerebellar pontine tumor, *labyrinthine disease* and chronic serous arachnoiditis is difficult. Barany's<sup>1</sup> method of accentuating symptoms by intra-aural caloric injections may be useful.

**Blastomycotic Meningitis.**—This chronic form of meningitis may closely simulate a tumor with no localizing symptoms. We have had two recent examples both mistaken for neoplasm, the cerebrospinal fluid in both having been normal aside from greatly increased tension.

**Hydrocephalus.**—It seems hardly possible that a tumor could be mistaken for chronic or essential hydrocephalus, but such is the case, not only in the presence of a slowly progressive cerebellar lesion, but even with large tumors arising above the tentorium which happen to obstruct the ventricular outflow. Diastasis of the sutures occurs early in the cerebellar tumors of the young, and the cranial enlargement is symmetrical: in tumors of the hemispheres an asymmetrical configuration of the head is more apt to occur, although even in these cases an associated ventricular hydrops may lead to a condition with a symptom-complex similar to simple hydrocephalus. A careful analysis of the incidents of onset will usually serve to distinguish the two conditions.

A low grade of choked disk, rare in simple hydrocephalus, is almost always present with the tumor cases; and the latter are more apt to present rigidities with exaggeration of the reflexes and ataxia of movement than are the former. In one of our cases diagnosed by a number of neurologists and surgeons as hydrocephalus consequent upon ependymitis, an enormous psammoma of the right hemisphere was found; another child treated by ourselves for a year as a simple hydrocephalic proved to have a cystic glioma of the cerebellum.

**Hematoma of the Dura Mater.**—Many conditions of lesser moment must be differentiated at times from tumor, such as hematoma of the dura

<sup>1</sup> Barany, *Wien. med. Woch.*, August 27, 1910.

mater, characterized by a neomembrane of inflammatory origin with increasing layers of bloody extravasation. This may be found to cover the entire convexity of one hemisphere or may be merely a local process. It is usually accompanied by symptoms of progressive general paralysis.

**Traumatic Cysts.**—Such cysts or even those of congenital origin may simulate tumors or be actually associated with tumors.

**Lead Encephalopathy.**—This, according to Bramwell,<sup>1</sup> may present headache, vomiting, convulsions, often epileptiform in character, and double optic neuritis, and must be carefully distinguished from tumor when there is any suspicion of lead poisoning. He affirms that the two conditions so closely resemble one another that he never commits himself to a positive diagnosis of intracranial tumor without previously excluding lead poisoning.

**Pseudo-tumors.**—Under this caption Nonne,<sup>2</sup> Oppenheim, and Hoppe have reported a number of instances in which a definite tumor symptom complex has completely subsided under specific or general medical treatment, and others in which, on postmortem examination, there was absolutely nothing to account for the definite pressure and focal symptoms present during life. Needless to say, hysterical conditions are excluded by the presence of definite symptoms of organic disease, often with a choked disk. We have had two cases of supposed tumor with death following operation in which no cerebral lesion was disclosed; and it is possible that a number of the patients in whom all evidences of pressure completely and permanently subsided after decompression have been cases of "pseudo-tumor cerebri."

Certain of the œdemas of one origin or another, meningitis serosa and like conditions, are very difficult to recognize after death, and they represent states which lend themselves favorably to operation. The surgical cases which recover after simple decompression, even when they have been of sufficiently long standing and severe enough to have caused optic atrophy from choked disk, may in some instances represent healed tubercle, abscess, or cystic metamorphosis of a glioma. Hoppe calls attention to the possibility that some of the pseudo-tumors with negative postmortem findings may be due to the rare chronic cerebritis or cerebral hypertrophy described by Rokitsansky.

**Course and Prognosis.**—Generally speaking, an intracranial tumor represents one of the most serious of maladies, dreaded by victim and physician alike, in view of the suffering to be endured by the one and the incapacity of the other to relieve. As the diagnosis is rarely certified until pressure symptoms have become full-blown, the duration of life from that time rarely exceeds six months or a year and is often preceded by mental deterioration and blindness. Speaking of the lesions in particular, on the other hand, in all their multiplicity of structure, manner of growth, situation, and possible complication, no definite statement can possibly be made. Their onset is always insidious, but it is self-evident that of tumors of the same size and type, one which occupies a silent area remains symptomatically dormant much longer than one which

<sup>1</sup> *Clin. Studies*, 1908-09, N. S., vii, 91.

<sup>2</sup> *Deut. Zeit. f. Nervenheilk.*, 1907, xxxiii, 317.

arises, for example, primarily in the precentral gyrus, or which early in its course has involved one of the cerebral nerves.

One thing is certain, that tumors are of much longer duration than is generally supposed, for the retrospect will often show that even malignant gliomata have doubtless been present for months or years before a diagnosis is made coincident with the appearance of general pressure symptoms, after which their course may be rapid. The symptoms from the onset are usually progressive, although there are notable exceptions, for tumors may enter a stationary phase and undergo retrograde metamorphosis. A tubercle may become quiescent, a glioma cystic or calcified; and some of the benign growths, as the endotheliomata, are of years' duration, leading in some instances to "autotrepanation" with relief of pressure, or they may even be a totally unexpected postmortem finding.

There are other exceptions to the usual rule according to which symptoms slowly progress from bad to worse. Thus in cases with sudden or apoplectiform onset or when symptoms come on abruptly after a fall the entire story is apt to be a short one. Then, too, death is not uncommonly abrupt and unexpected, owing to a sudden acute œdema or to medullary anemia from pressure, so that prognosis as to duration of life is most uncertain.

There remain "the unsolved riddles of pseudo-tumors," in the language of Bruns, so that the prognosis, in the presence even of a typical tumor symptom complex, need not necessarily be utterly pessimistic.

**Treatment.—Medicinal Measures.**—So little has treatment been able to offer that the physician has heretofore turned to the one possible therapeutic resource—antiluetic measures—on the chance that the lesion might be syphilitic. The length of time which can justly be given over to such treatment in a case of brain tumor has been much debated. The period required for definite results has been placed by some as high as six months, and the dosage has been enormous. In this interval or even in a much shorter one if the growth happens not to be luetic, the patient may rapidly fail and his choked disk, if one be present, go on to the atrophic stage. Even a person in health, put upon a similar treatment, would suffer greatly from nutritional disturbances, and much more the individual already upset by headache and vomiting.

Indeed, as a diagnostic test antisymphilitic treatment may be most misleading, for two reasons: one because with certain gliomata there may be a temporary amelioration of symptoms under these measures; and the other because the fibrous syphiloma, which gives the most characteristic tumor syndrome of all syphilitic processes, is exceedingly resistant even to massive doses of the usual drugs. Hence, even when the history and a positive serum reaction assure the diagnosis, the lesion must in the end be removed by operative methods if it be localizable, or if not, a palliative operation is necessary before the pressure symptoms are sufficiently relieved to allow the patient to receive full benefit from the drug administration. This too is the more urgently demanded in case a high grade of choked disk exists.

The typical basilar gummatous meningitis is relatively easy to diagnose and yields much more quickly to treatment than does a syphiloma.

Fortunately the Wassermann or Noguchi reaction will usually serve to spare these patients much discomfort and their attendants long anxiety. The tests should always be made at the earliest moment.

To state this matter briefly, the writer would say that in the presence or absence of a positive serum reaction or a definite history of lues, anti-syphilitic treatment deserves only a brief vigorous trial; if pressure symptoms have been outspoken and do not become distinctly ameliorated in the course of a few days, a palliative decompression should be performed and the treatment subsequently resumed should the diagnosis remain in doubt. One valuable criterion of the effect of the treatment and the need of early operation must rest with the condition of the eye-grounds, which should be observed daily, for a choked disk may continue to advance, even though subjective discomfort is lessened. In our series a fibro-syphiloma has been found and successfully extirpated in a number of instances in patients who had been subjected to a long although unavailing course of treatment and were much run down.

The general treatment should be directed toward the best possible preservation of the nutrition by frequent small amounts of digestible food; for, apart from the incidental vomiting many of these patients tend in later stages to lose weight rapidly. Cerebral congestion is to be avoided by proper position of the head, by ice caps, and by free daily evacuations. Alcohol is not tolerated. There is practically no effectual way of warding off the explosive attacks of vomiting; the headaches when insufferable may require large doses of narcotic drugs, antipyrine, phenacetine, codeine, combined with chloral, trional, or veronal for the night, and the hypodermic use of morphine as a last resort. Convulsions must be combated by bromides, or in desperate conditions by chloral or morphine—even by inhalation narcosis. Occasionally a threatened Jacksonian attack may be aborted by the prompt inhalation of a few drops of chloroform.

**Surgical Measures.**—The first therapeutic indication for a tumor anywhere in the body which occasions, or is likely to occasion, symptomatic disturbances, is *removal*, provided this can be accomplished without risk and without leaving undesirable consequences; for the mere prolongation of life, whether through medicines or operation, provided it is not made worth the living, is, of course, undesirable. The second indication is the *alleviation* of symptoms, and inasmuch as the conditions in the case of intracranial growths are almost entirely mechanical, they demand, for anything more than temporary palliation, mechanical means of relief.

The first ray of hope in regard to the operative therapy for brain tumors was dimmed by von Bergmann's widely heralded views as to the practical hopelessness of these measures, owing to his long series of early surgical failures. But within the past few years new elements have served to put the subject on an entirely new and much sounder basis. Among these have been the greater precision in diagnosis and a greater perfection in technical operative methods. A further element of no little significance is the development of surgeons trained in neurological pathology and diagnosis—in other words, of operating neurologists; for it is inconceivable that these matters of handicraft can advance far or fast when the surgeon

is merely the hand directed by the neurologist—a relationship comparable to that of the pre-Vesalian anatomist and his prosector.

If medicine is still to be widely separated from surgery a system of medicine is no place for a discussion of surgical procedures. Nevertheless, so long as the physician continues to be the one first called upon to determine the future course of treatment for a patient with brain tumor, it is the more needful that he shall understand something of the operative methods employed and the results which should be obtained thereby.

It is astonishing to learn how rapidly the number of these operations has increased. Allen Starr, in 1893, succeeded in collecting 97 cases; Chipault, in 1894, 135 cases; Oppenheim, in 1896, 140 cases; whereas today these by no means represent individual figures. Thus in the two years since the opening of the Brigham Hospital there have been 199 admissions for "Brain Tumors," and, in all but a few of these, operative procedures were employed which gave a promise of relief or cure.

Surgical measures have two main objects: (1) To expose and remove the growth if possible; (2) to palliate the symptoms of pressure in case (*a*) of an impossible regional diagnosis, (*b*) of a lesion in a recognizable though surgically inaccessible region, (*c*) of irremovability on exposure, whether from excessive size or undue vascularity.

**TUMOR EXTIRPATION.**—Practically all of the successful cases before the year 1900 were confined to tumors involving the paracentral convolutions, but during the past ten years the number of successful removals from frontal, temporal, parietal, or occipital lobes and from the cerebellum has rapidly increased. The exposure of the hemispheres is no longer carried out by simple trephining, but by the reflection of a large osteoplastic flap with its base hinging on the thin temporal region. Thus, a wide area of exposure is secured through which the necessary manipulations can safely be carried out. As the making of the flap itself, from loss of blood, may be something of an ordeal, a two-stage operation, as advocated by Horsley, is often advisable. The tumor, if exposed, should not be scooped out, but should be removed, if possible, by careful, slow dissection, with painstaking hemostasis. A bone-flap operation is undesirable in cerebellar cases where a bilateral suboccipital exposure with permanent removal of the bone is essential. Subtentorial tumors are often favorable for removal.

**TUMOR PALLIATION (DECOMPRESSIVE OPERATIONS).**—The unexpected relief which followed many of the earlier unsuccessful explorations gave birth to the idea of making a purposeful defect in the cranium and dura to give relief to pressure symptoms. Owing to the tendency of the brain under pressure to herniate through a defect and of the protruding portion to lose its function, it is desirable to make the opening over a silent area. When it is desirable to end a fruitless exploration of the hemisphere with a palliative decompression, the osteoplastic flap may be removed *in toto*, or the subtemporal area of bone alone removed. In the case of presumed cerebellar lesions the palliative operation differs in nowise from that necessary for exploration.

These successful palliative measures have in many ways represented our chief advance in the past few years, for they are comparatively simple

and free from risk. Their chief value lies in the prevention of blindness, which too often is the penalty of procrastination in cases of brain tumor. Certain lesions included under intracranial tumors, as well as actual tumors themselves, may prove to be more or less self-limited, with a tendency toward spontaneous healing, but unhappily loss of vision through optic atrophy is apt to occur in the interval of their activity; and to offset this danger, decompressive operations are particularly well adapted.

With the exception of tumors which have produced an internal hydrocephalus, alleviation of the pressure symptoms without increase in focal manifestations may be expected in most cases; and the subtemporal protrusion, if well protected by muscle, is very unobtrusive. Occasionally a callosal puncture (van Bramann) may serve to alleviate the symptoms of a secondary obstructive hydrocephalus. Tumors situated in the pontine region, with or without hydrocephalus, are also an exception to this general rule, for the change of position of the parts, either after the establishment of a subtemporal or a suboccipital defect, is apt to exaggerate preëxisting local symptoms.

Puncture of the brain (*Hirnpunktion*) and of the lumbar meninges as therapeutic measures, although successful in an occasional rare instance of draining an unsuspected meningitis serosa (Quinke) or the exceptional aspiration of a simple cerebral or cerebellar cyst, are nevertheless attended with grave risks.

Parts of the brain, like the interpeduncular region, heretofore thought forever secure from surgical approach, have proved to be accessible in a number of ways, and tumors involving the hypophysis, as well as primary hypertrophies of this gland, are regarded as surgical maladies.<sup>1</sup>

Notable advances have been made during the past decade in perfecting the technical methods of operations for brain tumor and the mortality of these procedures has fallen to a very low figure<sup>2</sup> compared with that of a few years ago. Nevertheless success in this particular field of surgery requires a special training the foundation of which must be laid in the neurological clinic and experimental laboratory for neuropathology.

<sup>1</sup> H. Cushing, *The Pituitary Body and its Disorders*, J. B. Lippincott Co., Phila., 1912; also *Surgical Experience with Pituitary Disorders*, *Jour. Am. Med. Assn.*, 1914, lxiii, 1515.

<sup>2</sup> *Concerning the Results of Operations for Brain Tumors*, *Jour. Am. Med. Assn.*, 1915, lxiv, 189.

## CHAPTER X.

### HYDROCEPHALUS.

By HARVEY CUSHING, M.D.

A PATHOLOGICAL increase in the amount of cerebrospinal fluid in the cranial chamber doubtless always represents a symptom of disease, not a disease of itself; although it must be confessed that pathology is at a loss to explain the undue accumulation of fluid in a certain group of cases, which, therefore, have been styled "essential" hydrocephalus. It may occur as an acute or chronic process, congenital or acquired, due to inflammatory or purely mechanical causes; and finally the hydrocephalus may be confined to the ventricular spaces or be found on the external surface of the brain.

As the cerebrospinal fluid, through retention or overproduction, is the factor chiefly concerned, some account of its source and physiological action is essential to such a crude understanding of the condition as we possess. Although possibly suspected by some of his predecessors, nevertheless Magendie, in 1842, first established beyond doubt the normal presence and rapid reformation of the fluid after withdrawal.

Notable recent additions to our knowledge have come, first, through the reawakening of interest in the fluid, as the result largely of Leonard Hill's physiological and Halliburton's clinical studies in the nineties, and second, through the introduction and widespread use of Quincke's lumbar puncture. Hill expressed the opinion that the fluid should be regarded as the lymph of the brain, but as Halliburton<sup>1</sup> showed, it has marked chemical differences from a transudation, and must be regarded as a true secretion, for the normal fluid is clear and colorless—like water—slightly alkaline in reaction, and of low specific gravity, 1005 to 1010; containing a mere trace of proteid (globulin) coagulable by heat, and also a reducing substance which has been shown to be glucose.<sup>2</sup> Lymph is like diluted blood plasma, with a specific gravity of 1012 to 1022, giving a test for sugar, containing proteids (albumin) in large amounts and coagulating spontaneously. Only in diseased conditions, such as inflammation and general paresis, does the usual character of the cerebrospinal fluid become greatly altered, with an increase of proteid, higher specific gravity, and in paresis a new substance, choline, due to the presence of degenerated nerve cells (Mott and Halliburton).

The fluid, in large part at least, is presumably the product of secretory activity of the ependymal cells lining the vascular choroid plexuses, although the precise function of these peculiar organs has not been

<sup>1</sup> *Biochemistry of Muscle and Nerve*, Philadelphia, 1904, p. 70.

<sup>2</sup> An elaborate monograph entitled *Le liquide céphalo-rachidien normal et pathologique*, Paris, 1912, W. Mestrezat, has fully considered the subject.

conclusively established. Certain experiments have shown that their cells may be definitely activated; and the plexuses doubtless possess an actual glandular function.<sup>1</sup> The secretion appears to be a fairly active and more or less continuous one, for the fluid, under certain circumstances, may form in large amounts. This is well illustrated by the rapid refilling of the hydrocephalic ventricles after tapping, but perhaps is best seen when, by accident or intent, an external fistula communicating with the subarachnoid space has been established, or in the remarkable cases of cerebrospinal rhinorrhœa such as are described in St. Clair Thomson's monograph—a condition characterized by a spontaneous escape of cerebrospinal fluid from the nose. No symptoms save loss of weight may attend such an escape of fluid continued over months, and an amount equal to half a liter may be collected in twenty-four hours. There may be a double source for the fluid, and although the larger portion comes through the ventricles, this may be added to by the cerebral lymphatics, which are said to follow the pial vessels and empty into the arachnoid spaces.

It would seem, therefore, that there must be a definite circulation for the fluid; and presumably forming in large part in the lateral ventricles, it passes thence by the foramen of Munro to the mid-ventricular system and escapes into the subarachnoid spaces by way of the foramina of Magendie and Luschka. From this point it bathes cord and brain, confined chiefly in the subarachnoid spaces, for only in certain pathological conditions does fluid appear in any considerable amount in the subdural space. From the subarachnoid spaces the fluid seems to escape directly into the circulation by way of the dural sinuses, rather than through lymphatic channels with the intermediation of glands before it reaches the venous system—another striking difference from true lymph. Some of it may possibly escape along the meningeal sheaths of the cerebral and spinal nerves—particularly the optic (cf. the condition in choked disk) and olfactory, but this is but a drop to the rapid escape by way of the sinuses. It was the view of Key and Retzius that the Pacchionian granulations played a certain rôle in this escape, but though it presumably occurs at the points where they are situated, fluid may escape freely, by way of the arachnoidal connections with the dura, in infants, in apes, or in the canine, in whom no Pacchionian granulations are present. Recent studies by Weed<sup>2</sup> made it clear that it is the arachnoid villi or tufts, which project into the dura, which represent the points of exit for the fluid.

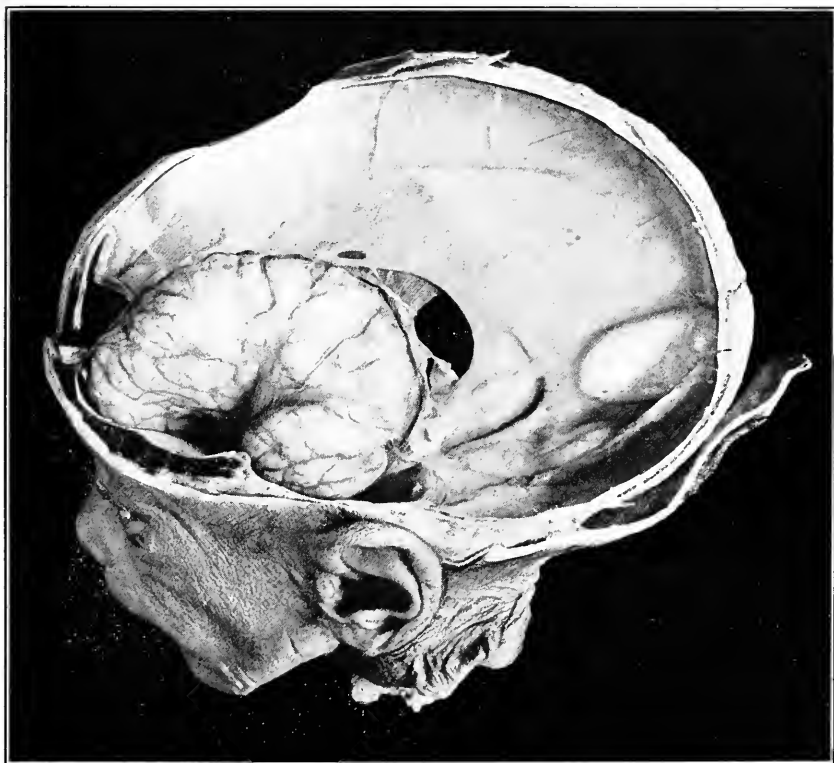
Even with our scant understanding of the cerebrospinal fluid circulation, it nevertheless can be seen how ventricular hydrops, the more important of the conditions we are to consider, can be due to obstruction of the channels of outlet from the ventricles; how infectious processes may easily occlude the small perforations in the roof of the fourth ventricle, or the arachnoidal points of entry of the fluid into the sinuses; how thrombosis of the sinuses may lead to cerebrospinal fluid stasis, apart

<sup>1</sup> Meek, *Jour. of Compar. Neurol. and Psych.*, 1907, xvii, 286.

<sup>2</sup> Studies on the Cerebrospinal Fluid, *Jour. of Med. Research*, 1914, xxxi, 1-176.



PLATE VI



Hydrocephalus Externus in an Infant with Normal Development of Skull and Meninges, but Defective Brain.



from the possible actual increase in the amount of fluid through venous congestion; how it is that the source of obstruction may be easily overlooked; and lastly, why many of the operations suggested for the relief of the condition have been based on erroneous principles of drainage.

**Hydrocephalus Externus.**—An abnormal collection of fluid outside of the ventricular cavities is rare, if we are to exclude certain pathological states accompanied by œdema of the brain and leptomeninges. It is misleading to refer to the “wet brain” of alcoholism, to the traumatic or angioneurotic œdemas, or to some of the cerebrospinal disturbances grouped under serous meningitis, as external hydrocephalus, although there is an excess of fluid in the meningeal spaces. It is confusing also to include here the *hydrops ex vacuo* or “compensation hydrocephalus,” which is merely the collection of fluid required to fill the intracranial chamber when atrophic processes have led to a shrinkage of the brain.

A true external hydrocephalus, however, may be found in association with congenital anomalies, when an undeveloped brain is found filling, in part only, a cranial chamber of normal configuration (Plate VI); and in rare instances it may occur in combination with a ventricular hydrocephalus when, through spontaneous perforation, the fluid has escaped into the subdural space. This, however, on physical principles would not be expected to occur very often, and surgical experience has shown that the distended and thinned hemisphere will not necessarily collapse and allow fluid to become extracerebral, even if a large opening be made into the ventricle through its thinned wall.

**Hydrocephalus Internus.**—It is customary to distinguish several varieties, particularly the so-called *idiopathic* form, regarded as a *primary* hydrocephalus, from those in which the condition is palpably *secondary* to some manifest lesion—the *acquired* forms. It would seem, however, that if there is any form of hydrocephalus which may justly be considered as a primary hydrocephalus, it is represented by the acute collection of fluid associated with the meningitis serosa of Quincke or with ependymitis; for here it is possible that we may be dealing with a condition in which fluid is excreted in excess of what may readily escape from the cranial chamber by the normal outlets. Nevertheless the resultant thickenings of ependyma when the acute process has subsided often lead to a persistent hydrops, so that even these states are customarily included with acquired or secondary hydrocephalus.

**Essential or Idiopathic Hydrocephalus.**—**Etiology.**—This sometimes occurs as a familial type, several members of a family being afflicted, so that hereditary influences play a part. The condition has been ascribed to fetal syphilis or rachitis, its association with congenital lues having been frequently observed. It is deserving of note, however, that the lower animals, especially those in captivity, may have hydrocephalic offspring. Presumably in most cases there exists some obstacle to the normal outflow of fluid from ventricle or cranial chamber, whether due to anomaly or disease. Heinecke believes that the hydrops is produced by an abnormal increase in the amount of fluid secreted. Both factors possibly may be at work.

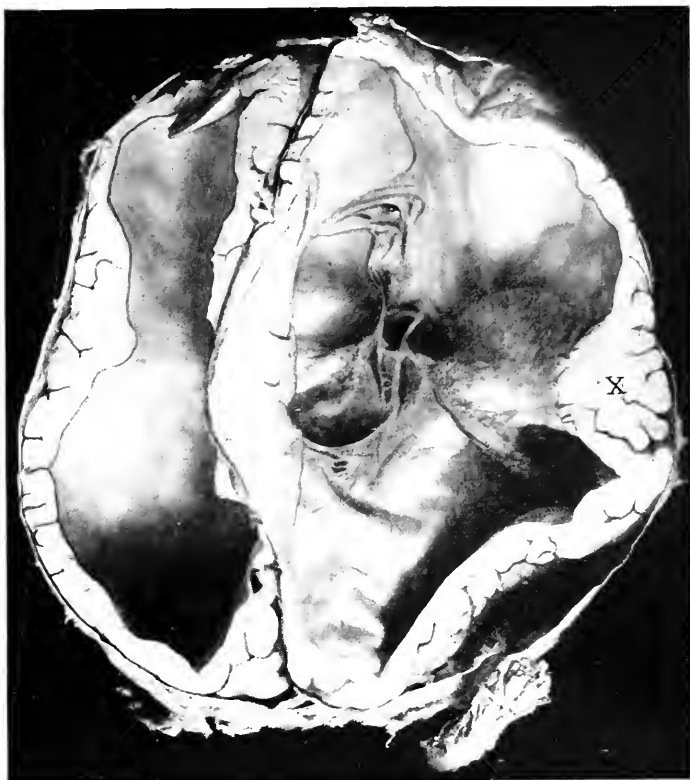
**Pathology.**—Meningeal thickenings suggestive of some prenatal inflammation are occasionally found matting together the structures, particularly about the roof of the fourth ventricle. More often, however, such an evident source of obstruction is absent, and the fluid from the ventricles can be withdrawn from the subarachnoid spaces by lumbar puncture, showing that if the process is actually an obstructive one the stasis in the fluid circulation must occur elsewhere, possibly at the foramina of entrance into the sinuses. The nature of these foramina has been made clear by Weed's studies and as the fluid escapes through the delicate membrane lining the arachnoid tufts by slow seepage an inflammatory process of mild degree may easily occlude these structures.

Ventricular hydrocephalus is the usual accompaniment of the structural anomalies, *cephalocele* and *spina bifida*, and it is a natural conjecture that these developmental defects have been produced by some early disturbance with the normal circulation of cerebrospinal fluid which may have prevented the proper closing in by the mesoblastic tissues of the primitive nervous system. This could easily be brought about by some abnormal increase in tension of the fluid before closure was complete. Only when the obstructive lesion, whatever it may have been, has been recovered from or overcome by some operative measure can the spina bifida or cephalocele be removed without considerable augmentation in the degree of hydrocephalus already present.

"The appearance of the skull is striking, the large, thin, flaring cranial leaflets being perched on the small facial bones like the petals of a single water lily on its calyx. The bones themselves are thinned and atrophied in places, so that pericranium and dura may meet (craniotabes). There is apt to be an abnormal number of Wormian bones. The frontal, temporal, and occipital wings, instead of arising vertically from the base, overhang so that the squamous wing of the temporal, for example, becomes almost horizontal and overlies the zygoma in such a way as to almost obliterate the temporal fossa. The irregularities of the three basal fossæ are slowly pressed out, so that the sphenoidal and petrosal ridges separating them no longer project as prominent watersheds. The base, however, does not participate in the general enlargement which the other bones undergo. In extreme cases the brain itself becomes ballooned out by the gradual increase of fluid, so that in places it is of paper thinness. Not only the ventricles, but the communicating passages also are all widely dilated; the iter is distended, and the foramen of Munro may become large enough to admit three fingers (Plate VII, Fig. 1). The corpus callosum may be stretched into a thin sheet and the much distended septum lucidum often gives way, leaving a direct communication between the lateral ventricles. The commissures between the basal ganglia usually remain intact, although they may be drawn out to a length of 2 cm. or more" (Plate VII, Fig. 2).

The amount of fluid may be enormous—three liters or more in extreme cases. It shows no abnormality in saline, albuminous, or cellular elements.

**Course and Symptoms.**—In its more familiar and striking form the condition has been evident from birth, or at least has set in with insidious



Advanced Grade of Chronic Internal Hydrocephalus, with extreme thinning of hemispheres, except at pyramidal tract (X). Note enormous size of foramen of Monro. Circumference of head, 70 cm.



Moderate Degree of Ventricular Distention in Chronic Hydrocephalus. Note rupture of septum lucidum; also great widening of iter, which is crossed by elongated commissure.



onset in early infancy. Owing to the ready distensibility of the skull at this early age the condition when full-blown leads to external appearances which are unmistakable; "a cranial chamber enlarged out of all proportion to body and face; the enormous and tense fontanelles; the cranial bones of the vault widely separated, even when they should be together, so that the midparietal, midfrontal, parieto-temporal and parieto-occipital sutures, bridged over by tense membrane, may all be open for 1 cm. or more; the bulging of the frontal temporal, and occipital bones; the thin scalp, with its sparse hair and dilated vessels, which are thrown into prominence, on account of the intracranial venous stasis; the expressional characteristics due to the tilting of the ears, with their wide intertragical notch and slitlike meatus; of the eyes, which are displaced by the downward bulging of the orbital roof and are covered chiefly by the lower lid, owing to the upward pull of the tight scalp upon the palpebral angles. The palpebral cleft is narrow, and the upper lid, when smoothed out, shows a network of dilated vessels."

When the enlargement is extreme (Plate VIII, Fig. 1) these patients are unable to turn or raise the head, and pressure decubitus may result unless changes of position are frequent. The circumference may often reach 70 and occasionally 100 cm. and the circumferential form is variable, depending upon the degree of tension and the habitual position assumed by the head, whether on the side, back, or at an angle.

The patients are, for the most part, feeble and poorly nourished, and even if the process becomes stationary after it has reached a high stage they have little resistance and are prone to succumb to intercurrent troubles. Should they live for some years they are apt to retain an infantile aspect, as well as infantile mental characteristics. Blindness, from optic atrophy, may occur, but this is not usual, and, indeed, even a low grade of retinal œdema is rare, for a pressure sufficient to produce an advanced degree of choked disk cannot well occur, as the cranium is so easily distensible. In some instances, even of the extreme type, adult life may be reached, with a certain feeble intellectual capacity combined with a traditional good nature. Rarely the process seems to have become arrested at an early age, with subsequent normal mental and physical development. Indeed, certain individuals with notable intellects are said to have shown evidences, post-mortem, of a low grade of hydrocephalus, presumably congenital.

**Diagnosis.**—There may be a close resemblance of a low-grade process to the rachitic head; the conditions of course may co-exist. When hydrocephalus is uncomplicated by this nutritional disturbance the diagnosis usually presents little difficulty. For though the cranium is large, cranio-tabes present, the fontanelles open, and the lines of the sutures flexible in rickets, the square shape of the head, the absence of signs of extra-cranial venous stasis, a fontanelle which is not tense, and rachitic changes in the bones elsewhere tend to distinguish the process. The chief difficulty lies in the differentiation of this so-called idiopathic hydrocephalus from the acquired form secondary to definite lesions. This is largely a matter of history; for in the essential form the process has been present from birth and is often associated with other anomalies; in the acquired form

Careful inquiry will show that a normal development has seemingly been interrupted in consequence of a cranial injury, a febrile illness, or some other unusual incident. A choked disk and the hydrocephalic rigidities are much more common in the acquired form.

**Treatment.**—The indications are purely mechanical ones and should be directed toward the establishment of some form of permanent drainage, whether by the channels intended by nature to carry away the fluid or by the establishment of new ones. There are a great number of operative procedures in which the attempt is made to drain the fluid into the subarachnoid space, into the subaponeurotic space of the scalp, directly into the sinuses, into the abdominal cavity, into the retroperitoneal spaces, and so on. None of them have given particularly brilliant or uniform results, although there have been occasional recoveries even in advanced cases. Whatever method is used, it is necessary to determine first of all where the obstruction has taken place, for many of the older operative measures are futile if the ventricular and subarachnoid spaces freely communicate. Most of the older operations have been conducted on the view that occlusion of the neighborhood of the foramen of Magendie is the single possible lesion. Temporary withdrawal of the fluid by ventricular or lumbar puncture leads, as a rule, to its rapid reaccumulation, at times under greater tension than before, and is rarely of any lasting benefit.

**Acquired or Secondary Hydrocephalus.**—This differs from the essential or primary form only in so far as it is apt to have a definite onset subsequent to birth, often in association with a manifest causal agency.

**Etiology.**—Any form of obstruction to the normal circulation of the fluid or any condition which increases its production in excess of what may escape through the normal channels of outflow will produce this form. The more common agencies are tumors, meningeal or ependymal inflammations, and stasis of the venous circulation through extensive sinus thrombosis, or obstruction of the *venæ Galeni*.

**TUMORS.**—Tumors, particularly those confined in the subtentorial region, whether intrapontine, extrapontine, or cerebellar, may obstruct the ventricular iter or the foramen at the roof of the fourth ventricle, leading to a more or less symmetrical dilatation of third and lateral ventricles. When these conditions arise in early infancy there is a rapid secondary enlargement of the head, so that the usual pressure phenomena of tumor may be wanting, with external appearances the exact counterpart of those described under essential hydrocephalus. A tumor, a cyst, or a tubercle may be an unsuspected postmortem finding (Plate VIII, Fig. 2). Diagnosis may be impossible without a local exploration—and not always then. A most valuable aid is the history of a previously normal state before symptoms began to appear.

The outward manifestations of idiopathic hydrocephalus, with separated sutures, wide fontanelles, etc., need not be limited to the secondary hydrocephalus of subtentorial lesions, for tumors of the cerebrum may occlude one or both foramina of Munro and produce a symmetrical cranial enlargement with general increase of pressure which serves to mask all focal symptoms. The tumors of themselves, under these circumstances of a yielding skull, may reach an enormous size.



# PLATE VIII

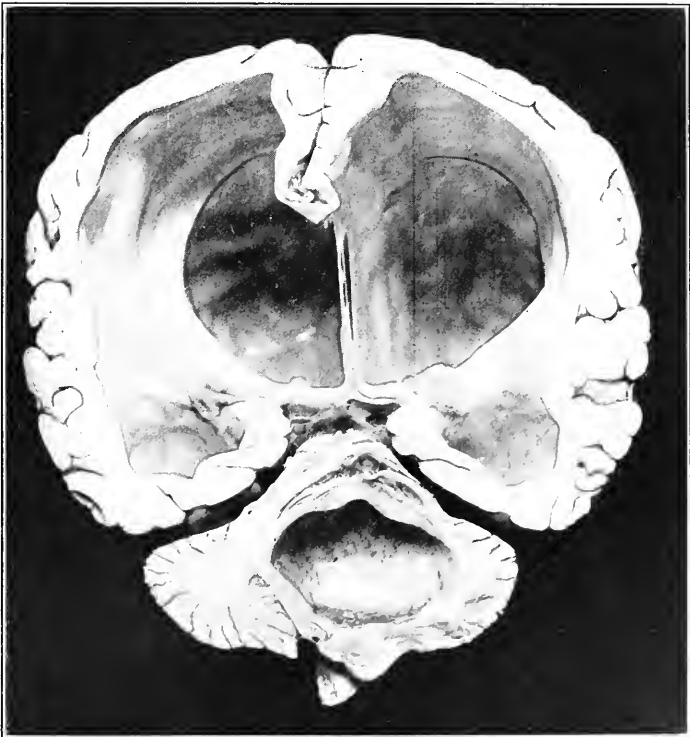
FIG. 1



Idiopathic Hydrocephalus.

An unusual degree of cranial enlargement. Circumference, 40 inches.  
(Dr. Willard Knowlton.)

FIG. 2



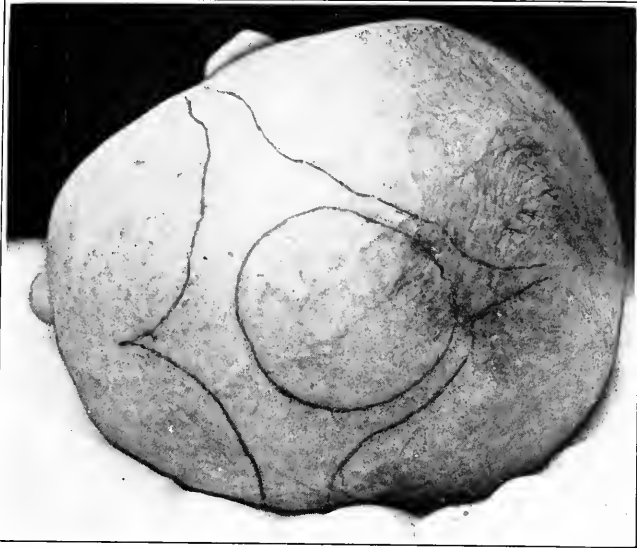
Unsuspected Gliomatous Cyst of Cerebellum in an Infant.

Producing enormous ventricular distention from obstruction; regarded during life as idiopathic hydrocephalus.



## PLATE IX

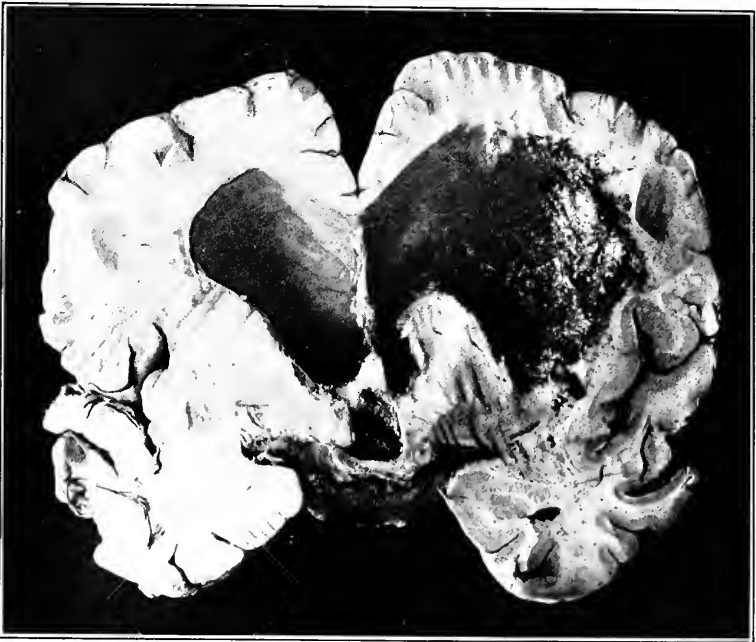
FIG. 1



Showing Separation of Sutures and Widely Opened Fontanelle Containing Isolated Island of Bone.

An infant with hydrocephalus secondary to meningitis; viable organisms in ventricles.

FIG. 2



Section of Brain of Fatal Case of Capsular Apoplexy.

In which a chronic hydrops ventriculorum with granular ependymitis was an unexpected postmortem finding.



Tumors which arise after the normal closure of the sutures and fontanelle has occurred may, through hydrocephalus, lead to a reseparation or diastasis of the sutures in the young, and even after adolescence, with an appreciable enlargement of the head, although never to such a degree as is seen in the infantile conditions. The "cracked-pot" percussion note which characterizes these conditions is an unmistakable evidence of a secondary hydrocephalus. Röntgenological changes in the skull are also very characteristic. The sutures are apt to show significant separations, but the most striking tell-tales of hydrocephalus are the pressure digitations or convolutional markings on the inner surface of the skull. There is only one other condition, namely, oxycephaly—which gives this picture in a degree more striking than hydrocephalus.

The abrupt onset of general pressure symptoms, in many cases of tumor which have been more or less dormant, is usually due to the occurrence of an obstructive hydrocephalus. This is particularly true of such benign growths as the cerebellopontine endotheliomata. The pressure symptoms are not necessarily constant, but may vary in intensity from time to time, depending on the degree of completeness of the obstruction.

**INFLAMMATIONS.**—A basilar inflammation, provided the exudate serves to occlude the minute foramina of exit in the roof of the fourth ventricle, leads inevitably to a hydrocephalus—the usual antecedent of death in these processes. Were it not for this mechanical obstruction and the resultant pressure phenomena meningitis would doubtless often be a self-limited disease.

**Cerebrospinal Fever.**—Attention has been called to the fact that in meningitis a pyo-hydrocephalus is often the terminal incident, for with the ventricular spaces not only infected but occluded the fatal termination of the malady is not remote. In infants, however, the ventricular distension need not give profound pressure symptoms, for the cranial chamber may rapidly enlarge (Plate IX, Fig. 1) and the process continue for months, organisms remaining viable in the cavities after they have died out from the meningeal spaces.<sup>1</sup> Naturally in these conditions serum introduced by the lumbar route does not have access to the infected surfaces, and introduction by ventricular puncture is indicated.

**Posterior Basic Meningitis.**—In the longer enduring forms of infection, as in Barlow's posterior basic meningitis, these occlusions are common, and the disease may drag on for months with slowly enlarging head, persistent retraction of the neck and hydrocephalic rigidities. Thus, it is not uncommon for the cases to run a chronic course, with more or less fluctuation of the symptoms according to the ease with which the pent-up fluid may escape. The occluded foramina of exit may become rechannelled in some cases, with no further advance in symptoms, although even with recovery a ventricular hydrops of this sort is apt to persist in some degree.

The ventricular complications of *tuberculous meningitis* differ nowise from those due to other infectious agencies. The patients usually die with, if not from, the secondary ventricular dilatation.

<sup>1</sup> Cushing and Sladen, *Jour. of Exper. Med.*, 1908, x, 548.

*Ependymal Inflammation.*—A primary ependymal inflammation may obstruct the passage of fluid by inflammatory closure of the interventricular foramina. This mysterious condition characterized by the presence of a granular thickening of the lining ventricular membrane occurs more often in the adult than in the young. The condition has no distinct symptomatology aside from that of an intracranial infection with definite pressure symptoms. It is not infrequently recovered from, judging from the number of instances in which traces of its former occurrence (a dilatation of the ventricle with ependymal granules) without history are unexpectedly brought to light when death has occurred from other causes (Plate IX, Fig. 2).

*Meningitis Serosa.*—The meningitis serosa of Quinke is a condition which lumbar puncture has enabled us to recognize. It is an acute process, with onset very similar to that of infective meningitis, but without the association of organisms—at least of those which can be cultivated. The condition may arise spontaneously or in the course of infectious diseases or intoxications. It is a particularly common sequel of otitis media and may be difficult to distinguish from infective meningitis or abscess. There is an excess of fluid under tension and usually a ventricular hydrocephalus. A choked disk of low degree is commonly present in this as well as in the many definitely obstructive forms of ventricular hydrocephalus. In the absence of fever tumor may be closely simulated. Lumbar puncture shows the fluid to be under great tension, and its withdrawal may completely change the clinical picture for the better.

## CHAPTER XI.

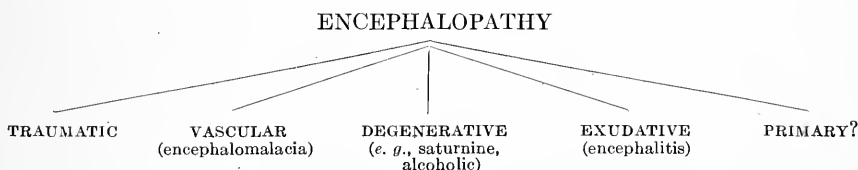
### ACUTE ENCEPHALITIS AND BRAIN ABSCESS.

By E. E. SOUTHARD, M.D.

#### ENCEPHALITIS.

**Synonyms.**—Inflammation of the brain; acute red softening of brain; brain fever; cerebral fever; cerebritis; phrenitis; phrenesia; phrenetiasis; phrenicula; cerebral phlegmasia; *calenture*; *febbre perniciosa frenetica*; *suppuratio occulta cerebri*. The terms brain fever, cerebral fever, and the like frequently refer to meningitis. The term encephalitis has been used freely in the past for entirely obscure conditions, so that I am assured by the United States Census Office that their figures for encephalitis are quite unreliable.

**Definition.**—Encephalitis means inflammation of the encephalon or brain. The best usage includes under the term encephalitis *exudative* inflammations of the brain. The place of encephalitis as here discussed may be indicated in the following scheme:



Most of the work on encephalitis preceding the era of Virchow's work on thrombosis and embolism (1846) can be neglected for modern purposes. Virchow's work began to have wide currency in 1856, but, although it cleared the way for a definition of focal softenings (focal encephalomalacia), the status of encephalitis was no better than before. Nor can it be said that Virchow's contribution of *encephalitis neonatorum* (1867)<sup>1</sup> aided systematic work, since it was at once opposed by Hayem<sup>2</sup> (1868) and Jastrowitz (1870), and cannot yet be said to be a solved problem. Hayem (1868) developed the idea of non-suppurative or hyperplastic encephalitis. Various contributions followed in which the infectious character of encephalitis became probable. Wernicke,<sup>3</sup> in 1881, described the disease (*poliencephalitis acuta hemorrhagica superior*), which bears his name, as an acute inflammatory process about the Sylvian aqueduct region, and compared the process with acute anterior poliomyelitis.

<sup>1</sup> *Arch. f. path. anat., etc.*, Berlin, 1867, xxxviii.

<sup>2</sup> *Etudes sur les diverses formes d'encéphalite*, Paris, 1868.

<sup>3</sup> *Lehrbuch der Gehirnkrankheiten*, Kassel, 1881, Band ii, Th. 3, S. 229 to 242.

Strümpell<sup>1</sup> (1884) constructed a very productive hypothesis—that many cases of cerebral palsy in children are due to encephalitis of the motor areas. In 1893 Macewen's work on *Pyogenic Infective Diseases of the Brain and Spinal Cord* appeared, in which the surgical importance of brain abscess and inflammation of otitic origin was developed. In 1898 Councilman, Mallory, and Wright described encephalitic lesions due to Weichselbaum's meningococcus, and in all cases co-existent with frank meningitis.

The infectious character of several types of exudative inflammation of the brain is well established. The interest of the present day is in the bacteriology (parasitology?) of these processes and in the possibility of differentiating cases into groups according to their infectious origins. Clinically, we are as yet unable to define groups with precision according to the different known infective agents. For some years, in Boston, an effort has been made to correlate these points of view. This work started out from the report of Councilman, Mallory, and Wright on "Cerebrospinal Meningitis," and has taken the shape of studies dealing with the histopathological and clinical effects upon the brain of the *Micrococcus aureus* (Southard and Sims,<sup>2</sup> Southard and Keene<sup>3</sup>), pneumococcus (Bullard and Sims,<sup>4</sup> Southard and Keene<sup>5</sup>), streptococcus (Southard and Stratton<sup>6</sup>), and *Bacillus typhosus* (Southard and Richards<sup>7</sup>). This work has shown the association of *Staphylococcus pyogenes aureus* and of the pneumococcus with acute hemorrhagic encephalitis, uncomplicated by meningitis.

**Classification.**—We distinguish the acute from the subacute and chronic forms of encephalitis. In the acute form we find characteristically the cardinal signs of inflammation. Of these signs, redness and swelling are prominent postmortem features of acutely inflamed brain tissue. The pain which sometimes accompanies encephalitis is due not to compression of the cerebral tissue itself, but to the pressure incidentally exerted upon the trigeminal endings by the swelling of the brain. Alterations of temperature can be demonstrated under surgical or experimental conditions only. More prominent and important than these signs is the so-called fifth sign of inflammation, or *functio laesa*, which may so affect consciousness at some stages as to remove the feature, pain, which is striking in inflammation elsewhere.

In the *subacute* and *chronic* forms we meet a variety of reactions which have, until recent years, been little understood. Both the histological findings and the clinical pictures in these latter forms have much in common with the degenerative class of encephalic disease. The terminal effect of repair in several classes of encephalitic disease (inflammatory, traumatic, degenerative) is atrophy or induration (sclerosis, gliosis), or even porencephaly, which may offer few or no features to determine

<sup>1</sup> *Jahrb. f. Kinderh.*, Leipzig, 1884, vol. xxii.

<sup>2</sup> *Jour. Am. Med. Assn.*, 1904, xliii, 789.

<sup>3</sup> *Am. Jour. Med. Sc.*, 1905, cxxix, 474.

<sup>4</sup> *Boston Med. and Surg. Jour.*, 1904, cli, 647.

<sup>5</sup> *Jour. Am. Med. Assn.*, 1906, xlv, 13.

<sup>6</sup> *Ibid.*, 1906, xlvii, 1271.

<sup>7</sup> *Jour. of Med. Research*, 1908, vol. xix.



the nature of the original disease. These points partly explain why there has been so much confusion as to what the term encephalitis precisely denotes.<sup>1</sup> Encephalitis means, strictly speaking, an inflammation of brain substance; but we habitually include also inflamed brain tissue in which there is little or no sign of parenchymal change and the chief sign of injury is perivascular infiltration. Within the brain substance, we might well distinguish between (and do so denominate certain diseases) *poli(o)-encephalitis* (inflammation of gray matter) and *leuk(o)-encephalitis* (inflammation of white matter).

*Chronologically*, we distinguish *acute encephalitis* (of which the types produced by staphylococci, streptococci, and pneumococci have been most studied), *subacute encephalitis* (of which the best examples are seen in syphilis and in tuberculous meningitis), *chronic encephalitis* (e. g., healed cases of epidemic meningitis), and *progressive encephalitis* as in dementia paralytica, now thought due to spirochætes.

Since the differentiation of thrombotic from inflammatory softenings, the development of entities in encephalitis has been as follows:

*Encephalitis congenita*, Virchow, 1867.

Spontaneous hyperplastic encephalitis, Hayem, 1868.

*Poliencephalitis acuta hemorrhagica superior*, Wernicke, 1881.

Acute infantile poliencephalitis (infantile cerebral palsy), Strümpell, 1884.

Acute primary encephalitis (adults), Strümpell, 1890.

Pyogenic infective disease of the brain, Macewen, 1893.

Encephalitis (*Bacillus influenzae*), Nauwerck, 1895.

Acute hemorrhagic encephalitis (*Staphylococcus pyogenes aureus*, pneumococcus), Southard and Keene, 1905 and 1906.

The most extensive experimental work on the pathogenesis has been by Friedmann. The most influential systematic treatise since Huguenin, 1876, has been Oppenheim's chapter in *Nothnagel's Handbuch*, 1897, now in its second edition (Oppenheim and Cassirer, 1907).

The picture of acute hemorrhagic encephalitis seems to have been produced by salvarsan (Fischer, Almkvist<sup>2</sup>) and even by mercury (Plötzl and Schüller).

The etiological basis has, so far as possible, been adopted in the classification which follows:

**Etiologically Well Characterized.**—*Influenzal Encephalitis. Pyogenic Encephalitis. (Hemorrhagic Superior Poliencephalitis, alcoholic form.)*

**Etiologically Doubtful, Clinically Definite.**—*Acute Hemorrhagic Encephalitis, Strümpell type*, often pyogenic (?), sometimes influenzal (?), or unknown in origin. *Acute Bulbar Myelitis. Acute Encephalomyelitis. Rabic Encephalitis* (not considered in this article).

**Etiologically and Clinically Obscure.**—*Acute Non-suppurative Encephalitis, Hayem type. Encephalitis in the Newborn.*

To these must be added other forms of such rare occurrence as to forbid systematization. Thus, *Bacillus typhosus* (as well as other

<sup>1</sup> Nové-Josserand, *Thèse de Lyon*, 1909–1910.

<sup>2</sup> *Münch. med. Wochenschr.*, 1011, lix, 1803, 1809.

typhaceæ), *Bacillus anthracis*, and *Trichinella spiralis* have been found in encephalitis. The encephalitis following scarlet fever, measles,<sup>1</sup> and other exanthems is probably of pyogenic origin. The relation of encephalitis to gonorrhœa, mumps, and whooping-cough is still doubtful etiologically.

**Influenzal Encephalitis.**—The great pandemic of influenza in 1889 to 1890 led a few writers to suspect the influenzal origin of encephalitis.<sup>2</sup> The influenza bacillus was cultivated by Nauwerck from a case of encephalitis in 1893–94. He found the bacilli in section from a soft hemorrhagic spot in the cerebellum and in the sediment of the ventricular fluid. The organisms were cultivated from the ventricular fluid. Trouillet and Esprit<sup>3</sup> published in the same year a report of pure cultures from the ventricular fluid in a disease which they term *Meningo-encéphalopathie grippale*. It was suggested that influenza toxins might prepare the tissues for other organisms, notably for the pneumococcus, which might be recovered in pure culture from the meninges or brain substance.

The atrium of infection in brain influenza may be: (1) The blood; (2) the nasal mucosa and the cribriform plate; and (3) the lymphatics of the middle ear. The nasal mucosa is now considered to be the most frequent route of infection, but it is probable that influenza is frequently a blood infection. Pfuhl<sup>4</sup> found three cases of influenzal encephalitis in an epidemic of 93 cases of influenza in the barracks at Hanover.

Encephalitis is not, as a rule, part and parcel of an attack of typical influenza, but follows recovery or partial recovery from such an attack. Sometimes there is an interval of several weeks before cerebral symptoms set in. In fact, the previous attack of influenza may be quite forgotten or considered as a cold in the head. Oppenheim remarks that the premonitory symptoms of influenzal encephalitis may be hard to interpret, since the headache, vertigo, nausea or vomiting, apathy, drowsiness, and general weakness may be regarded as merely incidents in convalescence from ordinary influenza. In other instances the onset may be acute.

If there is anything which distinguishes this form of encephalitis, it is a gradually increasing loss of consciousness, taking several hours or a day for its completion. The patient can, as a rule, be roused by sharp stimuli, and the pupillary reflexes are normal. The projection system is not characteristically involved at the onset. Meningitic symptoms, such as stiffness of neck and opisthotonos, are exceptional. There is irregularity in the temperature, which is rarely high until the cerebral symptoms are well under way. Some of the febrile temperatures are perhaps due to intercurrent infections (Oppenheim). But these statements were made before the question of influenzal septicemia had engaged attention. The pulse is likely to be slow at first, but before death quicker, small, and irregular.

The focal symptoms develop, as a rule, after coma has set in, not apoplectically, but one by one (Stadelmann's case with onset suggesting

<sup>1</sup> Marsh, *Brit. Jour. Children's Diseases*, London, 1910, vii, 20.

<sup>2</sup> Putnam, *On Multiple Neuritis, Encephalitis, and Meningitis after Influenza*, *Boston Med. and Surg. Jour.*, 1892, vol. cxxvii.

<sup>3</sup> *Sem. Méd.*, 1895, No. 21.

<sup>4</sup> *Zeit. f. Hyg. u. Inf.*, 1897, Band xxvi.

cerebral hemorrhage is a rare exception). Moreover, in a limb about to become paralyzed there are often premonitory symptoms, such as weakness, numbness, or convulsions. Sometimes the convulsions involve the whole side of the body, when the eventual loss of power is to be a monoplegia. The paralyzes may be quite masked by coma, or a difference in the two sides may be detected by choreiform movements on one side only. The variety of onset and course is so great that Leichtenstern felt that there are limiting cases in which a hemiplegia may develop without coma on a truly influenzal basis. Almost every possible focal symptom has been described in cases of encephalitis developing *after* influenza; but motor symptoms far outnumber sensory ones.

It was formerly thought that the prognosis was grave, but Oppenheim is inclined to agree with Leichtenstern and Fürbringer that many patients recover. The relation of influenzal encephalitis to spastic infantile hemiplegia cannot yet be stated; probably many of these cases are the relics of early coccal infections following exanthems.

**Pyogenic Encephalitis.**—Macewen states that pyemic abscess of the brain may arise from infective embolism originating from pneumonic areas, fetid bronchitis and empyema, fetid pericarditis, infective compound fractures, rarely acute infective periostitis, and occasionally infective ulcers of the intestines and abdominal cavity. In his experience abscesses were rarely multiple except when originating in pyemia. Macewen makes mention of purulent encephalitis, red softening, and white softening. He describes purulent encephalitis as consisting of a swelling of the cerebral substance with extensive serous and leukocytal exudation and extravasation of red blood corpuscles, which he recognizes to be of very frequent occurrence in acute inflammations of the brain. The coalescence of minute hemorrhages gave rise to red softening according to Macewen. He made the observation that the pressure occasioned by the œdema and swelling of the brain "may aid in producing the pain experienced by the patient at the very outset of the encephalitis, especially when the ganglia of the fifth and its recurrent meningeal branches are included in the area of pressure."

White softening is described by Macewen as due to infective embolism. He regarded microorganisms as the chief cause of intracranial pyogenic processes, and emphasizes the middle ear as an atrium of infection. He found the *Streptococcus pyogenes* and the *Staphylococcus aureus* most frequently in brain abscesses and in suppurative leptomeningitis.

In pyogenic encephalitis there is an invasion (whether primary or secondary) of pyogenic bacteria. Although the pyogenes in the broad sense include numerous bacterial species, in the writer's experience only the *Staphylococcus pyogenes aureus* and (in a few instances) the pneumococcus have produced pure examples of hemorrhagic encephalitis. Encephalitis complicated by meningitis or meningitis complicated by encephalitis (conditions usually united under the term meningo-encephalitis) is produced by the pneumococcus and streptococcus. In the writer's eight cases of pyogenic encephalitis the encephalitis was produced in six instances by *Staphylococcus pyogenes aureus* and in two instances by the pneumococcus. A possibility in the pathogenesis is an acute

inflammatory lesion of the arachnoid villi of the longitudinal sinus.<sup>1</sup> If this could be shown to be the rule, one reason could be assigned for the frequent affection of the central gyri in this disease.<sup>2</sup>

All the writer's cases of *Staphylococcus pyogenes aureus* brain infections have shown multiple hemorrhages in the brain. In two instances a condition which might be termed bacterial apoplexy has been produced in which rupture of a vessel and solution of surrounding brain tissue has been attended with intraventricular blood.

The pneumococcus, although cultivated somewhat more frequently from the brain and meninges than the staphylococcus, is less likely to produce hemorrhagic encephalitis.

In general, it may be stated that this disease occurs at all ages. Cases ranging from five to fifty years occur in the writer's series of pure hemorrhagic encephalitis; cases of meningo-encephalitis range from four months to ninety years of age. Alcohol is an occasional antecedent factor. In children the exanthems frequently precede encephalitis, but encephalitis may appear to be a late complication of an exanthem. Arterial disease appears to have little or no relation to pyogenic encephalitis. The onset is more apt to be acute than gradual. The cases of gradual origin in the writer's series were more apt to be in older patients. The course is short, although variable, ranging from twenty-eight hours in a fulminant pneumococcus case to two weeks. The diagnosis of hemorrhagic encephalitis was not made in the writer's cases. The symptoms pointed to lesions of the brain in all cases except two, in which diagnoses of septicemia and of acute disease of unknown origin were made. Meningitis was considered in two of the *Staphylococcus pyogenes* cases, and in one of these suspicion of poisoning was raised. In the cases of meningo-encephalitis the diagnosis of meningitis was frequent, and in 1 case a definite diagnosis of meningococcus meningitis was made. In 3 cases of meningo-encephalitis no other diagnosis than that of severe pneumonia was made. In the cases due to *Staphylococcus pyogenes aureus* the onset was sudden in 3 cases (hemiparesis and unilateral convulsions in 1; chilly feelings and frontal headache followed by ecchymoses in 1; malaise, manifold pains, ecchymoses, and cerebral symptoms in 1). In 1 case the symptoms gradually supervened upon a pleurisy and orbital suppuration.

The syndromes, as a whole, were of pronouncedly cerebral type in 2 cases, of meningitic trend in 2 cases, of septicemic character in 1 case. Septicemia could be suspected in 3 cases in which superficial ecchymoses or suppuration became prominent. The temperature varied for the various cases; at times 103° and 104° for 2 cases; the temperature in 3 cases was never high and frequently subnormal. The pulse was in no case subnormal, and, as a rule, ranged from above 100. The respirations were, as a rule, increased.

The duration of the encephalitic symptoms cannot, in all cases, be closely reckoned; in 1 case three days; 1 case one week (after development of erysipelatoid condition) or three weeks (after injury to eye); 1 case, eight days; 1 case two weeks; 1 case fifteen days; 1 case unknown. The

<sup>1</sup> Southard and Sims, *Jour. Am. Med. Assn.*, 1904, xliii, 789.

<sup>2</sup> Grasset et Rauzier, *Maladies du système nerveux*, 1894.

cases in which the cerebral syndromes were most clearly marked were of longer duration.

The distribution of the brain lesions was as follows: Four cases showed areas of frank and somewhat voluminous hemorrhage, as a rule involving primarily the subcortical region (the vertex in 1, the occipital lobes in 2, and in 1 the pons). Three cases showed multiple ecchymoses and small abscesses of the cortex or subcortical region, with or without gross evidence of surrounding œdema. In 3 cases œdema of the pia mater was noted. In 2 cases an effusion of blood into the lateral ventricles was noted; in these cases the convolutions were flattened. In 2 cases, in which the temporal lobe was involved, the middle ear showed no lesion.

A remarkable association of encephalitis with focal pulmonary lesions was made out in this series. Pulmonary lesions were either grossly prominent or found microscopically in all six cases. It is possible that this association is a mere matter of coincidence.

**Hemorrhagic Superior Poliencephalitis (Wernicke's Disease; Total Ophthalmoplegia; Nuclear Ocular Palsy; Alcoholic Ophthalmoplegia).**—First described by Gayet, 1875,<sup>1</sup> as *affection encéphalique (encéphalite diffuse probable)*, localized in the upper parts of the cerebral peduncles and in the optic thalami, as well as in the floor of the fourth ventricle and in the lateral walls of the third ventricle, the disease owes its name to Wernicke, whose cases were published in his *Lehrbuch* in 1881. The entity was further strengthened by Boedeker, who in 1892 summed up the findings in 11 cases (3 apparently non-alcoholic, and 8 alcoholic). The resemblance of the disease to Korsakoff's polyneuritic psychosis was brought out by Bonhoeffer (1901).<sup>2</sup> Schröder has shown that the disease, although a definite entity, is not a true encephalitis in the exudative sense.

Wernicke defined the disease as an acute inflammatory disease of the nuclei of the eye muscle nerves, leading to death in from ten to fourteen days. The disease was characterized by palsy of eye muscles of rapid onset, quickly progressing to almost complete ophthalmoplegia (sphincter iridis and levator palpebrarum excepted). The patient's gait recalled that of alcoholism, showing a combination of stiffness and ataxia. Somnolence was a striking feature, either at onset or toward the close. Where somnolence set in late, the disease at first recalled delirium tremens. Optic neuritis occurred in all Wernicke's cases. The course is not always so acute, for Gayet's case lasted five months, and other writers<sup>3</sup> described cases with recovery.

(a) *Hemorrhagic Superior Poliencephalitis, Non-alcoholic.*—These cases are very few. In Gayet's case the symptoms followed three days after a boiler explosion, in which, however, the patient, a man, aged twenty-eight years, sustained no bodily injury. Paraphasia and paraphagia were the first symptoms. General weakness, apathy, and marked *drowsiness* followed. The face became mask-like, and there was double ptosis with a general atony of the facial musculature, so that in speech there were no expressive facial movements. General weakness was such that the

<sup>1</sup> *Archives de Physiologie*, 1875, vol. vii.

<sup>2</sup> *Monatsschr. f. Psych. u. Neur.*, 1899, v, 265.

<sup>3</sup> Wolfe, *Jour. of Nerv. and Ment. Dis.*, 1894; Wiener, *Brain*, 1897, xx, 458.

patient could scarcely stand or grasp objects. Pupils, vision, and all kinds of sensation were quite normal. There was almost complete *paralysis* of both *oculomotor nerves*. There was later transitory hemiplegia, which disappeared, leaving the patient in a state of excitement, with more facial expression and pain in the right leg. Conditions varied thereafter. Intelligence was maintained, and there were never any considerable changes in the eye grounds. Emaciation followed with incontinence and decubitus; death occurred five months after the accident.

Morbid sleepiness also characterized Wernicke's first case, a young girl, who had attempted suicide with sulphuric acid. The sleepiness, ataxia, optic disturbances, and vertigo came on two months after the suicidal attempt. Oculomotor palsy, disorientation, and hebetude were the main features of the attack, which led to death twelve days after the onset. Salomonsohn's case showed loss of knee-jerks and slowing of the pulse, in addition to other characteristic symptoms. Luce's case was incidental in miliary tuberculosis. Murawieff's case followed three months after influenza, and showed also sarcoma of one precentral gyrus.

(b) *Hemorrhagic Superior Poliencephalitis, Alcoholic*.—This is closely allied in symptomatology with Korsakoff's disease and with alcoholic polyneuritis. Some writers believe that the symptoms in the three conditions scarcely represent entities, but are all incidents in chronic alcoholism. If this be true, it is doubtful whether Wernicke's disease is truly encephalitis in the exudative sense, and, in fact, Schröder was able to find in a typical case, merely, (a) destructive lesions due to minute hemorrhages, and (b) reparative changes (phagocytic cells, vascular and neuroglial changes) like those following any non-inflammatory injury. Schröder inclines to the belief that alcoholism sets up some kind of vascular change such that, under conditions of unknown character involving severe brain injury, capillary hemorrhages ensue in various parts of the brain. The region beneath the corpora quadrigemina is the locus of election for such hemorrhages. It is possible that this tendency is related to the characteristic hemorrhages in anaphylaxis.

Subjects having this symptom complex are often brought to a hospital as cases of delirium tremens. The victims are usually male whisky drinkers of middle age, who have been suffering for months or years from chronic alcoholism. For some days or weeks, headache, pains in various parts of the body, vertigo, and vomiting may precede the ophthalmoplegia. As a rule, the ophthalmoplegia is incidental in a characteristic alcoholic delirium, which otherwise differs from the ordinary delirium tremens in the well-marked mental and bodily weakness of the patient.

The ophthalmoplegia is not absolute nor always an associated palsy. Sometimes the pupil and the levator palpebrae superioris are involved as well. Nystagmus is frequent. Wilbrand and Sanger<sup>1</sup> pointed out that bilateral ptosis occurs in about one-half the cases, although it is not an essential feature. One case in six shows normal vision and unaltered eye grounds. The eye ground changes when found, consist either of a

<sup>1</sup> *Die Neurologie des Auges, Handbuch*, 1900, vol. i.

slight neuritis with retinal hemorrhages, or of a pallor of the temporal halves of the papillæ (sign of an alcoholic amblyopia due to chronic neuritis of the papillomacular fibre bundle of the optic nerve). It is difficult to distinguish the rest of the picture from that in chronic alcoholism. *Muscular weakness*, indistinct *schlaftrunken* (Wernicke) speech and a *gait* recalling the cerebellar gait are characteristic.

In the differential *diagnosis* brain tumor (especially of the posterior cranial fossa, particularly in the cerebellum) is suggested by the gait and the eye-ground changes. The course is perhaps more likely to suggest apoplexy or focal encephalomalacia. Not only the gait and the non-febrile course, but also the alterations of consciousness may support such a diagnosis; and, in the light of Schröder's histological observations, the disease may perhaps be best regarded as a peculiar kind of apoplectic disease in which multiple minute hemorrhages affect a particular area. The optic neuritis and the symmetry of the ophthalmoplegia eliminate any question of the classical forms of apoplexy. The ocular palsies of botulism (bilateral paralysis or paresis of accommodation, mydriasis, ptosis) present some resemblances to those of Wernicke's disease.

**Acute Hemorrhagic Encephalitis, Strümpell Type.**—It is probable that many of the cases of acute hemorrhagic encephalitis which Strümpell predicted in 1884 and described in 1890 are of pyogenic origin. Strümpell's first cases showed at autopsy a bilateral bronchopneumonia and a markedly enlarged spleen, in addition to the encephalitis. Cultures from the brain lesions proved negative. Strümpell, after dismissing the hypothesis that his cases belonged to the epidemic meningitis group, assumed that there must be a specific agent for what he conceived was a primary local infection.

It seems justifiable to put all primary cases of unknown etiology provisionally into this group, which further investigation will undoubtedly resolve. The newer ideas about septicemia have so altered our point of view that one naturally hesitates to classify any encephalitis as primary, on the ground that it may result from a blood infection. Since Landsteiner and Popper, and Flexner have successfully transferred anterior poliomyelitis from man to monkeys, there is a possibility that not only anterior poliomyelitis, but also acute encephalitis and Landry's paralysis, may be due either to an ultramicroscopic organism or to protozoa. Noguchi reports successful cultivation of minute organisms.

Orazio D'Allocco described six cases of acute poli-encephalitis in children (one with autopsy). The onset in these cases was sudden, attended with general infectious symptoms. There was a continuous generalized tonic spasm of all the muscles, including those of respiration and of the head, with continual uniform almost rhythmical spasms. The limbs were affected symmetrically, as a rule, but the spasm may be confined to one limb. Spasms are more marked near the head. Muscular excitability is increased. Anatomically, an endophlebitis and an endarteritis leading to vascular occlusion and degenerative or inflammatory lesions were found. Henning, 1914, brings evidence of four groups of atypical cases, (a) epileptiform, (b) pseudotumor, (c) resembling multiple sclerosis, (d) psychotic.

**Acute Bulbar Myelitis (Acute Inferior Poliencephalitis; Acute Inflammatory Bulbar Palsy).**—*Acute inferior poliencephalitis* has more verbal than substantial resemblance to *acute superior poliencephalitis*, since the non-inflammatory hemorrhagic character of the latter (Schröder) is replaced in the inferior form by acute inflammatory lesions. These lesions recall closely those of anterior poliomyelitis; in fact, many cases of anterior poliomyelitis show bulbar involvement, so that some writers believe that acute inferior poliencephalitis is a disease closely allied to anterior poliomyelitis—possibly an identical infection with an unusual localization. W. Pasteur's<sup>1</sup> epidemic of seven cases of paralysis occurring in children (ranging from eighteen months to eleven years of age) in one family is often cited in this connection, since some of the cases were cerebral and some spinal. At all events, whether the analogy of anterior poliomyelitis is accepted or the frequent association of bulbar myelitis with influenza epidemics be taken as a hint at the etiology, the disease acute bulbar myelitis is very different from Wernicke's disease both in site and in character. Histologically, there is evidence of acute destruction of nerve tissue, not at all confined to the gray matter, together with vascular congestion and lymphocyte accumulations in the perivascular sheaths. Not only do these lesions affect white matter as well as gray, but the meninges are frequently involved.

The *onset* is rapid, but not apoplectiform. General malaise, headache, vomiting, and sometimes fever precede the local signs of bulbar involvement. The characteristic delirium or sleepiness of Wernicke's disease is not found. As a rule, in from three to seven days there is difficulty in swallowing, with paralysis of the tongue and palate, facial paralysis, and paralysis or paresis of vocal cords.

The course depends upon the locus of the inflammatory foci, which may directly involve a greater or smaller number of the gray nuclei (nuclear form of palsy), in which there is least hope of recovery, or may affect the nerves in their intrabulbar course (infranuclear form), in which the prognosis is less grave. On account of the ill-defined limits of the inflammatory process, any given case is somewhat likely at its height to show a combination of nuclear, infranuclear, and even supranuclear involvement. If the subject does not become unconscious, the course is said to be more favorable. The prognosis is not so unfavorable as was formerly thought; death occurs in about one-half the cases. The possible relation of certain cases of recovery to multiple disseminated sclerosis has been mentioned (Oppenheim).

In differential *diagnosis*, syphilis, hemorrhage, and softening of the bulb must be strongly considered, whereas tumor, tubercle, and abscess are less likely to cloud the issue. Syphilis is ordinarily attended by basal meningitic symptoms. Hemorrhage and softening have a sudden or rapid onset which is not characteristic of encephalitis. Some cases with persistent bulbar phenomena have been attributed to encephalitis; thus, for example, Huet and Lejonne<sup>2</sup> attribute a facial palsy and hemiatrophy

<sup>1</sup> *Clin. Soc. Trans.*, London, 1897, xxx, 143.

<sup>2</sup> *Rev. Neurol.*, Paris, 1906, xiv, 105.



of the tongue to the effects of a former acute inferior poliencephalitis attacking the nuclei of the facialis and hypoglossus of the right side.

**Acute Encephalomyelitis and Poliencephalomyelitis.**—These names have sometimes been given to cases of anterior poliomyelitis complicated by lesions higher than the spinal cord; but there are other cases in which the element of anterior poliomyelitis is not so prominent. Possibly cases of Wernicke's disease with a number of hemorrhages below the level of the quadrigemina have yielded so complex a picture as to deserve this name. Cases of acute bulbar myelitis have been not frequently complicated by lesion higher than the bulb. A tendency of the symptoms to *descend, possibly beginning with ophthalmoplegia*, going on with bulbar symptoms, and ending with limb disorders, has been noted. There is great irregularity in the degree to which the various portions of the involved apparatus are diseased. Whereas total ophthalmoplegia and glossopharyngolabial palsy, associated with spinal palsy, are the symptoms of the disease in its full development, yet in cases in which the ophthalmoplegia is complete the bulbar trouble may be limited to slight facial palsy; and in cases in which the glossopharyngolabial palsy is complete there may be no more than ptosis to express the suprabulbar lesion.

When the cerebral white matter and the included nerve paths are involved, it seems better to name the disease *encephalomyelitis*. The disease has foci so widespread in some instances as to lose its usual purely motor character (Taylor<sup>1</sup> and Oppenheim). These cases vary extremely in outcome, some dying, some recovering, and some remaining with defects. The probable course of several weeks or longer should possibly align them rather with the slower degenerative processes than with acute encephalitis. General cerebral symptoms are few or confined to the drowsiness and weakness characteristic of Wernicke's disease. Fever sometimes occurs. The facies is said to be characteristic (Hutchinsonian; eyeballs immobile, ptosis, elevated eyebrows), together with relaxation of the lower face, flattening of nasolabial folds, loose-hung limbs.

A case of Combe's<sup>2</sup> illustrates the peculiar combinations of symptoms possible in encephalitis with multiple foci. It is said to have followed influenza. That form of alternate paralysis known as *Weber's syndrome*, viz., right hemiparesis and left ophthalmoplegia, appeared in a boy, aged two years. Weir Mitchell's *posthemiplegic choreiform movements* followed on the right side, but associated with spasmodic movements in the field of the left facial and abducent nerves, amounting to an alternating hemichorea. Later there was weakness of the right oculomotor nerve. Such hemiparesis with crossed paralysis of the oculomotor nerve with tremors of the paralyzed parts corresponds with *Benedikt's syndrome* (due to a lesion of the cerebral peduncle). But the involvement of the left facial and abducent nerves, together with the right-sided hemiparesis, amounts to the *Millard-Gubler syndrome*. The patient, therefore, suffered from a combination of Benedikt's syndrome and the Millard-Gubler syndrome due to a lesion of the pons near the left pyramidal tract (this new syndrome Combe proposes to term the *Inferior Benedikt syndrome*),

<sup>1</sup> *Boston Med. and Surg. Jour.*, 1903, cxlviii, 634.

<sup>2</sup> *Syndrome de Benedikt inférieur*, *Rev. Mens. d. mal. de l'enfance*, 1904, I, xxii, 1.

but in addition showed a complication by incomplete ophthalmoplegia, due to a separate lesion situated in the oculomotor nuclei.

The diagnosis of such multiple circumscribed lesions and the determination of their encephalitic nature is a matter for the future. At present the general diagnosis must be made largely by elimination of vascular and other focal possibilities and on the ground of the course of the disease. The poli-encephalitis here mentioned evidently has considerable alliance with the degenerative diseases of these loci. E. W. Taylor distinguishes the following in this field: Encephalitis, encephalomyelitis, acute ophthalmoplegia, acute superior and inferior poli-encephalitis, acute or apopleciform bulbar myelitis, poli-encephalomyelitis, and poliomyelitis.

It may be mentioned at this point that Batten<sup>1</sup> proposes to group anterior poliomyelitis with encephalitis, and neglecting the preëmption of the term "superior poli-encephalitis," or Wernicke's disease, Batten classifies as follows: Acute poli-encephalitis superior, including cases with frontal, central, occipital, and cerebellar localization; acute poli-encephalitis inferior, including cases in which cranial nerve nuclei are affected; acute poliomyelitis anterior. Batten regards both brain and cord disease as due to thrombosis of fine terminal vessels due to an unknown organism or toxin. Batten's classification is not yet proved, although there are some cases in which there is a thrombosis of the fine terminal vessels (a prominent feature in pneumococcus and streptococcus brain infections). It is not proved that thrombosis plays any part in the disease anterior poliomyelitis.

**Acute Non-suppurative Encephalitis (Acute Hyperplastic Encephalitis, Hayem).**—Among non-hemorrhagic forms, Hayem's hyperplastic type is least well recognized and is probably very rare, except in the sense that hyperplastic or proliferative phenomena are the invariable concomitants of all not too severe destructive injuries in the central nervous system.

**Encephalitis in the Newborn (Encephalitis Congenita; Encephalitis Interstitialis Congenita; Encephalitis Neonatorum; Stéatose Interstitielle Diffuse de l'Encéphale).**<sup>2</sup>—This occurs at times as the result of septic infection of gastro-intestinal or pulmonary origin (Fischl). A case is on record in which a pathogenic strain of *Staphylococcus pyogenes albus* was grown from the focal lesions (Fischl). Encephalitis may complicate the picture of syphilis in the newborn, although spirochætes are seldom demonstrable in the nervous system, even in congenital syphilis.

Aside from pyogenic, syphilitic, or other recognized forms of encephalitis in the newborn, there is another form, the *encephalitis congenita* of Virchow, which is still *sub judice*. Virchow believed that he had learned the cause of death in a considerable proportion of stillborn and newborn infants, attributing it to encephalitis and myelitis of congenital origin. The only analogies for this form of encephalitis were findings in the brains of children of variolous and syphilitic mothers. In general, he regarded congenital encephalitis as a brain disease analogous with the parenchymatous changes of the liver and kidney in the exanthemata.

Hayem, 1868, promptly criticised Virchow's findings, noting the

<sup>1</sup> *Lancet*, 1902, vol. ii.

<sup>2</sup> *Archives de Physiologie*, 1868, vol. i.

extreme frequency of fat-laden cells in the brains of the newborn. In the same year Parrot published a piece of work, since largely neglected, on what he termed diffuse interstitial steatosis of the encephalon, and offered clinical histories which proved to Parrot's satisfaction that these changes were due to lack of food. Parrot's results were reprinted without marked alteration in his monograph on athrepsia in 1877.<sup>1</sup>

Jastrowitz regarded the fatty cells figured by Virchow and by Parrot as incidental in the development of the brain. From an extensive material (65 cases), Jastrowitz stated that these cells are found in small numbers in the third month of fetal life, in greater numbers in later months, but as a universal finding in the cerebral white matter only in the newborn. The locus of election for the granule cells is the white matter, and especially the corpus callosum and its processes. The occurrence of groups of granule cells outside these usual loci, *e. g.*, in the gray matter of the cortex or in the basal ganglia, was regarded by Jastrowitz as pathological. The pathological fat-cell groups were thought by Jastrowitz to be due rather to infections and various maternal diseases than to developmental errors or atrophy (Parrot).

Modern opinion inclines to the view that the focal form, in which not only masses of fat-laden cells, but also injury of nerve fibres, can be demonstrated is the true congenital encephalitis. A case of the writer's has shown how streptococci may bring about in the newborn a reaction of the mononuclear cells rather than of the as yet ill-developed polynuclear cells in the meninges. Possibly this finding explains some of the cellular peculiarities of congenital encephalitis.

*Summary.*—There is a form of encephalitis in the newborn which is at times a cause of stillbirth or of death soon after birth, and is due to various factors (sometimes to pyogenic infection, sometimes to syphilis, and sometimes to unknown intoxications), perhaps more often maternal than non-maternal. The disease is histologically characterized by focal lesions of the cerebral white matter (sometimes visible in the gross) showing nerve fibre alterations and accumulations of fat-containing cells. Protracted cases may yield a neurological picture of irregularly distributed convulsions and pareses, respiratory rather than cardiac disorder, and initial, not continued, fever.

**Experimental Encephalitis.**—The field of experimental encephalitis has not been thoroughly worked. Coen<sup>2</sup> demonstrated mitotic figures in the nerve cells surrounding the region of necrosis. Friedmann described varying phenomena of experimental encephalitis, which in 1904 he summed up by saying that "former distinctions of encephalitis into 'forms' are unnecessary," since sufficiently strong stimuli (meningococcus, influenza bacilli, embolism, aseptic trauma, corrosive substances or heat) can bring about all the different conditions which have been termed encephalitis. In the more severe cases, according to Friedmann, all histological combinations of encephalitic processes may be found ranging from small-cell infiltrations to areas of softening with granule cell deposits

<sup>1</sup> *Clinique des nouveau-nés. L'Athrepsie. Leçons recueillies par le Dr. Troisier, Paris, 1877.*

<sup>2</sup> *Ziegler's Beitr. z. pathol. Anat., 1887, Band ii.*

and extensive areas of proliferation with giant cells. Various stages and degrees of these processes have been hemorrhagic, parenchymatous, and hyperplastic encephalitis.

After small traumatic lesions fibrillary sclerosis may follow; after more extensive lesions cellular proliferation is prominent. In cases of hematogenous origin, Friedmann emphasizes a primary infection of the small bloodvessels. These have been shown by Southard and Keene, and Southard and Stratton, to characterize pneumococcus and streptococcus infections of the brain. Koeppen<sup>1</sup> described one end result as a kind of *état criblé*, in which the fibrillar gliosis has been insufficient to fill the space of the destroyed tissue. In other cases a dense sclerosis follows.

Experimental work upon bacterial encephalitis scarcely admits systematization. Work upon the peripheral nerve roots and the method of invasion of the central nervous system by bacteria has been done by Homen and Laitinen, and by Orr and Rows.<sup>2</sup> It is evident from this work that more attention must be paid to the lymphatic route of possible infection. The writer and associated workers have produced experimental brain infections in a variety of ways, using several varieties of pyogenic bacteria. They have produced meningitis, encephalitis, and ependymitis, but so far neither a pure encephalitis nor the important lesions of the small vessels found in many cases of adult encephalitis has been produced in the guinea-pig. The time relations of the experimental brain infections in the guinea-pig are: *Staphylococcus pyogenes aureus*, intrapulmonary injections, after six hours, focal polynuclear exudation in the meninges; after twelve hours, ependymitis; after twenty-four hours, slight polynuclear leukocytosis in the meninges; after forty-eight hours, subependymal leukocytosis and moderate encephalitis (leukocytes in apposition with nerve cells). After three days meningitis, encephalitis, and ependymitis at their height; after four and five days lymphocytosis and phagocytosis; after six days, phagocytosis for leukocytes; after nine days very slight polynuclear leukocytosis, slight evidence of phagocytosis. After one month the tissues are negative, except for slight fibrillar gliosis, pigment-bearing phagocytes, lymphoid and plasma cells, and eosinophilic polynuclear leukocytes.

In work upon the *pneumococcus*, postorbital inoculations were employed. Many strains of pneumococcus gave no lesion. Polynuclear leukocytosis was noted as early as six hours. The exudate is at its height in three, four, or five days, and is largely confined to the meninges. Some of the strains produced lesions in which mononuclear cells were more prominent than polynuclear cells. Work with the *streptococcus* produced results analogous to those with the pneumococcus.

Southard and Richards investigated guinea-pig brain tissues after postorbital inoculation of *Bacillus typhosus*. In contradistinction to the early crisis of the aureus brain inflammations (three days) and the pneumococcus inflammations (four days), it was found that the *Bacillus typhosus* produced a critical phase from the fifth to the seventh day.

<sup>1</sup> *Arch. f. Psych.*, 1898, Band xxx.

<sup>2</sup> *Brain*, 1904, p. 460; *Review Neurol. Psych.*, 1906, iv, 25; *Brit. Med. Jour.*, 1907, i, 987; *Review Neurol. Psych.*, 1907, v, 345.

Mononuclear elements appear in the pial meshes in the typhoid inoculations by the seventh day. By the tenth day fresh exudative cells have ceased to appear. By the fourteenth day the disease has virtually passed and traces only of the old exudate are found.

Special work has been done by Cantani on the effect of influenza bacilli on the central nervous system of rabbits. He was able to kill rabbits by brain injection of influenza bacillus in proper amount and of proper virulence. The first symptoms appeared nine to ten hours after injection. Paraplegia of crural type was gradually followed by paralysis of all extremities and convulsions. The autopsies showed evidence of severe intoxication throughout the trunk, as well as meningitis, encephalitis, and ependymitis, with demonstrable influenza bacillus. Injections of killed bacilli into the brain in larger amounts produced the same results.

In connection with experimental encephalitis more work is desirable in animals. Encephalitis in horses has been long recognized; but it is commonly held that equine encephalitis has a very manifold etiology.

**Diagnosis.**—The diagnosis of acute encephalitis is not often successfully made, and often not made at all. Perhaps the diagnosis is rendered more accurately in children's clinics than elsewhere, owing to the interest attaching to the Strümpell type. Nevertheless, in the majority of cases the course is either so acute as to offer little chance of diagnosis, or else is such as to warrant a diagnosis of "cerebral disease." In a large number of the writer's series of cases of encephalitis and meningo-encephalitis the diagnosis of cerebral disease was made without attempt to differentiate between hemorrhagic encephalitis, massive cerebral hemorrhage, meningitis, and brain syphilis. The attempt of Oppenheim to systematize encephalitis has been so successful that the diagnosis will undoubtedly be made more frequently in the future.

There are few practical difficulties connected with the diagnosis of *hemorrhagic superior poli-encephalitis, alcoholic form*, since the history of alcoholism, the acute onset, the associated ocular palsy, the somnolence, delirium, the general weakness (and, as a rule, optic neuritis), stamp the disease. Although Oppenheim sought to reduce hemorrhagic superior poli-encephalitis, alcoholic form; hemorrhagic encephalitis, Strümpell type; and influenzal encephalitis to a single entity on clinical grounds, it does not appear that this position is warranted. Bonhoeffer's clinical claims and Schröder's histological observations go far to show that Wernicke's disease is a rare but perfectly definite disease.

Although Wernicke's disease presents few or no difficulties in diagnosis, the *Strümpell type of acute hemorrhagic encephalitis*, the form termed in this article *pyogenic encephalitis*, and *influenzal encephalitis* give considerable difficulty. The endeavor should be made in all doubtful acute "cerebral" diseases to establish or eliminate an influenzal origin. Influenzal encephalitis, it must be remembered, occurs *after* rather than during an attack of influenza. Cases occurring in epidemics are more frequently diagnosed than sporadic cases. Lumbar puncture proves of considerable aid. The data of Cohen's work on *septicemic meningitis* must also be considered.<sup>1</sup>

<sup>1</sup> *Annales de l'Institut Pasteur*, 1909.

Since many of the cases may turn out to be of pyogenic origin and of pyemic nature, it is probable that blood cultures will aid extremely in the diagnosis. The etiological data of cultures from lumbar puncture fluid and from the blood will not point unequivocally to encephalitis. On theoretical grounds the positive lumbar puncture culture will point rather to meningitis or to meningo-encephalitis than to a pure encephalitis. Close attention must be paid to all the clinical signs. If we group together influenzal encephalitis, pyogenic encephalitis, and hemorrhagic encephalitis of unknown origin (termed in this article the Strümpell type), the chief conditions to be considered in differential diagnosis are: (1) Other intracranial infective diseases, such as meningococcus meningitis, tuberculous meningitis, pyogenic meningitis, and sinus thrombosis; (2) brain tumor and brain syphilis; (3) cerebral hemorrhage and focal encephalomalacia; (4) certain conditions of cerebral intoxication resembling meningitis but anatomically negative; and (5) rarely acute poisoning, hysteria, and other diseases.

The differential diagnosis in the first group, *intracranial infective diseases*, is greatly aided by lumbar puncture with examination of the cells and bacteriological examination. This, in doubtful cases, should include an attempt to grow the influenza bacillus on blood media. The finding of blood in the lumbar puncture fluid may suggest chronic internal hemorrhagic pachymeningitis and cerebral hemorrhage as well as acute hemorrhagic encephalitis. Nor will every case of hemorrhagic encephalitis show blood or leukocytes (Lucas and Southard) in the cerebrospinal fluid. A careful cytological examination should be performed. Examinations for the tubercle bacillus or meningococcus and other pyogenic bacteria should be made. A lumbar puncture fluid without leukocytes, blood, or bacteria must not be considered as excluding the possibility of hemorrhagic encephalitis.

Stiff neck, retraction of the abdomen, and hyperesthesia are not common in encephalitis. Herpes has not been reported. Paralysis of eye muscles is exceptional. Nevertheless, on account of the extensive variation in foci affected by the multiple lesions of hemorrhagic encephalitis, it is improbable that there is any sign of meningitis which may not also occur in some cases of hemorrhagic encephalitis. The diagnosis frequently depends on the association of a monoplegia or hemiplegia with successive involvement of the paralyzed parts or aphasia coming on early. Similar phenomena in meningitis are frequently complications or sequelæ rather than central factors. The acute onset, the drowsiness or tendency to coma, vomiting, convulsions, and optic neuritis offer no points in differential diagnosis against meningitis, nor can much weight be placed upon the condition of the pulse or respiration.

As against *sinus thrombosis* the stasis of blood in the veins of the face has been mentioned by Macewen, but this is not always present. Sinus thrombosis runs a very acute course. The differentiation often fails, except in middle-ear disease.

With respect to the differential diagnosis from *brain tumor*, the onset, the fever, and rapid course are important. Hemorrhagic encephalitis rarely shows an altogether characteristic tumor syndrome. Oppenheim

especially warns against the immediate diagnosis of brain tumor in cases showing attacks of cortical epilepsy. Similar remarks apply to brain syphilis when it takes the form of gumma. Brain *syphilis* in the form of chronic meningitis also runs a quite different course from that of typical hemorrhagic encephalitis. Those rare cases of brain syphilis in which the disease is essentially an encephalitis must give rise to difficulty, except when a history of syphilis can be established. Since these cases occur in secondary syphilis, the differential diagnosis should not prove difficult.

Cerebral *hemorrhage* and focal encephalomalacia were formerly frequently diagnosed in cases of hemorrhagic encephalitis. In young persons with healthy cardiovascular systems the diagnosis of cerebral hemorrhage or thrombosis may not come into question; but, since hemorrhagic encephalitis occurs in older persons, there is a possibility of confusion. The cases termed by the writer *bacterial apoplexy* occurred in subjects twelve and twenty-two years of age. In such cases the differential diagnosis between hemorrhagic encephalitis and cerebral hemorrhage would be impossible on theoretical grounds. Practically the paralyses which characterize hemorrhagic encephalitis do not occur simultaneously with the onset of drowsiness or coma. The signs of severe cerebral disease are obvious long before paralyses or convulsions have set in. The paralyses of hemorrhagic encephalitis may increase in intensity from time to time in the course of several hours or a day, presumably owing to the gradual development of inflammatory lesions with oozing of blood. The absence of fever and of other signs of sepsis will indicate rather a cerebral hemorrhage or focal encephalomalacia due to arterial disease.

Cerebral *intoxication without anatomical signs* (the so-called pseudo-meningitis of Krannhals<sup>1</sup>) gives rise to the diagnosis of encephalitis, although more often to the diagnosis of meningitis in some cases. Krannhals' cases occurred in the influenza pandemic of 1889 to 1890. The occurrence of focal symptoms would ordinarily exclude this diagnosis. Poisoning was considered in one of the writer's cases (*Staphylococcus pyogenes aureus*) of bacterial apoplexy, which ran an acute course of three days. Attention to the acute infective feature of encephalitis will usually exclude the diagnosis of *hysteria* with certainty. *Uremia* and *acetonemia* may need to be considered.

Flachs<sup>2</sup> sums up points in differential diagnosis as follows:

*Encephalitis:* Localizing symptoms (monoplegia, hemiplegia, aphasia), tendency to recovery.

*Meningitis:* Retraction of abdomen; hyperesthesia; eye-muscle paralysis, persistent rigidity of neck.

*Tumor:* Headache, optic neuritis, slowed pulse, slow course.

*Sinus Thrombosis:* Stasis phenomena, acute course.

*Pseudo-meningitis:* Severe symptoms; high fever with nerve symptoms.

**Encephalitis in Advanced Years.**—There is neither predisposition nor lack of predisposition to hemorrhagic encephalitis in old age. The diagnosis as against focal encephalomalacia or cerebral hemorrhage is naturally difficult, especially since febrile conditions (bronchopneumonia)

<sup>1</sup> *Deut. Arch. f. klin. Med.*, 1895, liv, 89.

<sup>2</sup> *Fortschr. d. Med.*, 1910, xxviii.

may apparently lend infective features to focal encephalomalacia. Hoppe-Seyler<sup>1</sup> mentioned hemorrhagic encephalitis as a rare complication of pneumonia in the aged, and Siemerling, in the same *Lehrbuch*, mentions difficulty in differential diagnosis between sinus thrombosis and encephalitis. Lhermitte has described a "paraplegic dementia" on the basis of chronic changes, apparently degenerative and not exudative.<sup>2</sup>

**Encephalitis in Infancy and Childhood.**—The possible medicolegal significance of congenital interstitial encephalitis was mentioned by Virchow. Possibly this question should be raised more often in cases of "overlying." Pediatrics practically distinguishes acute encephalitis in infants and a form of encephalitis in older children which shows more tendency to recovery. Acute and frequently fatal encephalitis of infants is regarded as a primary encephalitis with origin unknown except in certain pyogenic cases. These cases show high fever, stupor or coma, convulsions at the onset as well as later, superficial breathing with occasional Cheyne-Stokes episodes, a rapid, feeble pulse, and sometimes bulging of the fontanelles. The neck and limbs are stiff and flexed. Strabismus sometimes occurs. The occurrence of slight paresis or monospasm of a limb or of the facial nerve is characteristic. The endeavor should be made to determine or eliminate a pyogenic origin. Lumbar puncture findings are helpful as against a meningococcus meningitis.

The encephalitis of older children is often secondary to exanthemata, diphtheria, or pertussis, and has an insidious origin sometimes suggestive of brain tumor. Recovery is the rule in these cases, which sometimes precede the development of frank epilepsy. The child is immobile except for tetanic spasm; the neck is stiff, and opisthotonos may suggest meningitis. Tremor of the limbs, facial palsy, monoplegia, aphasia, convergence or conjugate deviation of the eyes, and optic neuritis may occur. James Taylor<sup>3</sup> has given valuable data, and distinguishes the following varieties of infantile cerebral palsy:

Hemiplegic forms, resulting from encephalitis, vascular lesions and injury.	{ Acute encephalitis, infantile hemiplegia, infantile monoplegia, idiocy with cranial asymmetry, choreiform hemiparesis, hemiathetosis.
Diplegic forms, resulting from death of cortical cells.	{ Generalized rigidity, idiocy with general rigidity, paraplegic rigidity, spastic facial diplegia, amaurotic family idiocy, bilateral athetosis, choreiform diplegia, epileptiform myoclonus.

Only the diseases under hemiplegic forms are considered by Taylor to be encephalitic in origin, and of these, infantile (cortical) hemiplegia is the most important. The conditions found, in order of frequency, are acute encephalitis, thrombosis (arterial or venous), hemorrhage, and embolism. Alzheimer called attention to the relation between idiocy and encephalitis. According to Taylor the victims of infantile hemiplegia

<sup>1</sup> *Schwalbe's Lehrbuch der Greisenkrankheiten*, 1909, p. 236.

<sup>2</sup> *Sem. Méd.*, Paris, 1910, xxx, No. 50, 585.

<sup>3</sup> *Paralysis and Other Diseases of the Nervous System in Childhood and Early Life*, 1905.



are healthy and without hereditary stigmata at onset, and undergo between the ages of one month and six years severe symptoms, consisting of fever, convulsion, and often vomiting and coma. After a period varying from a day to a week the attack becomes less severe, but hemiplegia comes on first in the face, then in the arms, and lastly in the leg. This hemiplegia is flaccid for a few days and afterward spastic. The paralysis clears up to a considerable extent, but, as a rule, not completely. Such are the primary cases. A second group is incidental in certain specific fevers, and probably corresponds to the group of pyogenic encephalitides. Taylor holds that thrombosis is found in certain cases, and calls attention to the possibility of embolism from endocarditis.

Obstetrical injury or other forms of trauma and mechanical rupture of vessels in paroxysms of whooping-cough or in convulsions and meningeal hemorrhage occurring at birth are less important factors. The prenatal cases of infantile hemiplegia are likely to be of thrombotic origin as a result of maternal syphilis. The intrapartum cases are due to obstetrical injury, and the postnatal cases are in general encephalitic.

According to Taylor the following anatomical changes have been found in cases of infantile hemiplegia: Circumscribed atrophic sclerosis (in cases where the nerve elements perish, but the neuroglia survives), cyst formation (in cases where both nerve elements and neuroglia are destroyed), superficial shrunken patches resembling wet wash leather with adjacent atrophic sclerosis (of thrombotic origin), and porencephaly. The neurological phenomena of surviving cases (persistent facial paresis, stunting of growth of arm, choreiform and athetotic movements of arm, characteristic brachial contracture, distortion of bones and joints, post-hemiplegic disorders of voluntary and involuntary movements, trophic disturbances and arrest of development, and epileptic attacks) will not be more particularly considered here. About one-half of all cases of infantile hemiplegia are said to be subject to recurrent attacks of epilepsy which begin long after the onset of the hemiplegia.

**Prognosis.**—W. P. Lucas and the writer have studied the outcome in 29 cases of clinically probable encephalitis, finding 3 cases of epilepsy, which developed after an apparently normal interval. Over one-quarter of our cases showed mild but distinct evidence of mental defect, and a number of others showed "nervousness." Strabismus, developing in the disease, has persisted in 2 of the 29 cases. A special study of the hereditary relations of the psychopathic subjects does *not* show that they have a higher index of taint than the rest of the population.

**Treatment.**—The febrile nature of hemorrhagic encephalitis indicates certain general measures. Although the hemorrhages are, as a rule, of an oozing character, and the progress of focal symptoms depends rather upon spread of inflammatory lesions than upon massive hemorrhages, it is desirable to avoid any drug treatment which may stimulate the heart. The patient should rest in bed with the head higher than the trunk, and be shielded from light and noise. In the somnolent or comatose condition especial attention must be paid to the administration of food and the evacuation of the bladder and intestine. Incontinence occasionally occurs. Cold compresses or ice bags may be applied to the head. Except

in cases of chlorosis, venesection or leeches in the temporal or mastoid region are said to have given good results (Oppenheim). Drug treatment is unsatisfactory. Restless patients may require various narcotics or moist packs. Oppenheim warns against surgical treatment and even against lumbar puncture. It appears that there have been few surgical attempts at alleviation of this disease. It might seem that, were accurate localizing diagnosis possible in certain cases of massive hemorrhage, surgery would be helpful. In general, surgeons are doubtful of the value of surgical intervention in encephalitis. Operation in meningo-encephalitis (Macewen's case) has been done, and Oppenheim calls attention to Hahn's case (1896), in which trephining was performed under the impression that abscess or hemorrhage of traumatic origin was present.

Since in the future our diagnoses of encephalitis must improve in the line of etiology, it is possible that especially in the influenzal and pyogenic groups the possibilities of immunization<sup>1</sup> and opsonic treatment may be exploited. The somewhat rare chance of a syphilitic encephalitis must be considered.<sup>2</sup>

### BRAIN ABSCESS.

Brain abscess is a focal destructive disease of brain tissues, characterized by the presence of pus, and is ordinarily distinguished from tubercle, gumma, neoplasm, and cyst of softening, despite the fact that cavities with puriform contents may be found in these conditions. As in the rest of the body, artificial aseptic abscesses can be produced by chemical or physical means within the cranium. Non-experimental abscesses are now generally stated to be invariably due to microorganisms. It is useful to bear in mind, that even in man a variety of destructive aseptic lesions can be produced by violence from falls or blows, and that only months or years later these quiescent lesions may be occupied by bacteria. Knowledge concerning the frequency of bacteriemia giving no clinical sign has explained the otherwise mysterious occurrence of latent traumatic abscess.

**Etiology.**—A great variety of organisms has been demonstrated in brain abscess, not all of which are surely concerned in the original production of the lesion, but may settle in preëxistent lesions. Malinowski, 1891, showed that suppurative processes could sometimes be induced in the normal brain of the dog by injections of pyogens, but constantly in brains that had been injured ten days previously.

**Pathology and Pathogenesis.**—Abscesses of the brain occur in all sizes from approximately half the size of a hemisphere down to the barely visible. The smallest abscesses are, as a rule, pyemic in origin and multiple; they intergrade with encephalitis, the non-suppurative form of which may be so named merely because no pus is visible, whereas, polynuclear leukocytes may be easily demonstrable microscopically. Nevertheless, there is good reason to distinguish such interstitial inflammatory conditions from frank abscess, as in other organs and tissues. The larger abscesses are usually single and are frequently objects of surprise at

<sup>1</sup> Watson, *Lancet*, London, 1912, i, 1469 (influenza vaccine).

<sup>2</sup> Laache, *Ztschr. f. Klin. Med.*, 1911, lxxii, 203.

autopsy. The larger single abscesses are apt to be subcortical rather than cortical, and are likely to break through to the surface or into the ventricles late in the disease.

The pathogenesis is by extension from *external infected wounds* (acute traumatic abscess), by extension from *sinus thrombosis*, by extension through *diseased bone* or by *lymphatic* paths (otitic, rhinogenic, orbital), by *blood infection* (pyemic abscesses, abscesses metastatic from bronchiectasis, empyema). The pathogenesis of latent brain abscess is dubious, although there can be no doubt of the part played by blood-borne infection. Occasionally there is an actual demonstration of pigment derived from lung lesions in the metastatic cases.

The abscesses contain, as a rule, a green or yellowish-green, or yellowish pus, generally watery, and in long-standing cases sometimes almost clear. The abscess wall after some days (five or six, according to Friedmann's experimental data, eight or ten days in man) begins to show a membrane. This membrane shows a large amount of collagenous tissue, intermingled with neuroglia in the outer zones. The development of mesodermal reaction in these abscesses is greater than in most other intraneuraxial lesions (solitary tubercle excepted), but for all that there is no tendency to obliteration of the cavity. Such obliteration must be the rarest of events. The investing membrane may be so dense and coherent that the whole abscess may be stripped out, especially from a brain somewhat softened by postmortem change. Frequently death, from rupture into the ventricles with generalized pressure symptoms or rupture into the meninges with a terminal leptomeningitis, intervenes before such membrane formation.

**Symptoms.**—The general symptoms are those of heightened intracranial pressure, and in the main resemble those of brain tumor. Deeply lying abscesses tend to heighten intracranial pressure more than superficial abscesses. Subtentorial abscesses produce a general heightening of pressure by means of hydrocephalus. The general symptoms are:

1. *Headache.* It is rare for a case of brain abscess to run a complete course without headache. Even in the latent period of chronic abscess headache may occur periodically. The onset of fever, attacks of coughing and sneezing, the use of alcohol and coffee are likely to increase the pain. The headache is apt to be of the dull, bursting type, and is inclined to localize itself upon the side of the abscess. Cerebellar abscesses yield, as a rule, a regionary headache, but there is a surprising number of cerebellar cases with frontal headache. A certain number of cases, perhaps the majority, show localized sensitiveness upon pressure or percussion over the seat of the abscess. During the period of encapsulation the headache of abscess is perhaps less severe than that of tumor; but rapidly spreading abscesses yield headaches of maximal intensity.

2. *Vomiting.* This occurs in practically all cases, as a rule at the time when headache is most marked, without relation to the condition of the stomach, and often apparently excited by a change of position. Subtentorial abscesses are perhaps more prone to show this symptom.

3. *Vertigo.* Vertigo, unlike headache and vomiting, is inconstant in brain abscess, and perhaps characterizes only a minority of cases. More-

over, since the majority of cases with abscess symptoms give at least a suspicion of being otitic in origin, the vertigo runs a chance of confusion with aural phenomena.

4. *Slowing of Pulse.* At the height of the disease the pulse is usually subnormal (perhaps 50 to 56). Pulse rates of 40 and 30 occur, and rarely it may diminish to 10. A slow pulse is especially helpful in the differentiation of brain abscess and local ear disease and sinus thrombosis.

5. *Respiratory disorder*, usually with tendency to slowing, is not infrequent, particularly in cerebellar abscesses.

6. *Eye-ground Changes.* These are less common and severe in brain abscess than in tumor. They are found, as a rule, when the other symptoms of brain abscess are pronounced.

Generalized *convulsions* occur in a minority of the cases. Cerebral abscesses in children appear to cause convulsions more often than such abscesses in adults. Late in the disease and in cases in which the pus bursts into the ventricles convulsions are more common. Hebetude, sleepiness, indolence, and an appearance of fatigue are characteristic. Absolute coma occurs late and may occur repeatedly in a given case. Episodic attacks of restlessness, confusion, or excitement may set in at any time and are characteristic of the initial stage in cases having an acute onset. They are apt to accompany attacks of fever. Cases suggestive of the psychiatric diagnosis of delirium are rare. The latent cases may suggest melancholia.

*Temperature.*—Normal temperature may persist throughout, and periods of subnormal temperature characterize some cases. The initial stages of cases having acute onset usually exhibit fever. As Oppenheim suggests, the insidious onset of many cases and the subsidence of fever before the physician is called in many others have led to the idea that brain abscess is characteristically non-febrile. Naturally the ventricular and meningeal complications of the terminal stages may lead to fever. Macewen's data show that the majority of cases throughout observation show normal or subnormal temperatures. Perhaps the latent encapsulated abscesses of a chronic type are those which most characteristically fail to show fever. Certainly a persistent high temperature or a typically intermittent fever is not characteristic of uncomplicated brain abscess, that is, abscess without meningitis or otitis.

**Focal Symptoms.**—Although cases sometimes fail to yield focal symptoms, yet the majority yield fairly characteristic symptoms which often betray the nature and origin of the lesions. Thus the occurrence of motor symptoms is most characteristic in traumatic and metastatic abscesses; whereas frontal abscesses are characteristically rhinogenic; temporal and cerebellar, otogenic. A few metastatic and traumatic abscesses have been found localized in the occipital region. Abscesses of the pons, the medulla, and the brachium conjunctivum are decidedly uncommon. Local variations in the relatively normal tissue surrounding abscesses are commonly stated to account for rapid modifications in the symptom picture and the immediate diminution in symptoms following surgical drainage of abscess cavities.

1. *Focal Motor Symptoms.* These are: (a) *Epileptic symptoms* of the Jacksonian type whose distribution may easily serve to indicate the spread of the abscess. These phenomena naturally occur more often with the cortical than with deeper lesions. The possibility is to be borne in mind that the convulsions are due not to destructive cortical lesions, but to encephalitic or meningitic changes overlying abscess. (b) *Paralytic symptoms* tend to be monoplegic, and, if hemiplegia eventuates, the parts are successively and not simultaneously involved. *Disorders of sensation* are often masked by the patient's mental state.

2. *Frontal Symptoms.* These are frequently absent or hard to differentiate. The vicinity of the motor zone permits, in the midst of indeterminate symptoms, the development in later stages of brachial or facial symptoms. Conjugate deviation of head and eyes toward the side of lesion has been noted in connection with epileptic attacks. The mental symptoms which one might expect and which occasionally arise in the brain tumors so localized have generally failed of demonstration.

3. *Temporal Symptoms.* Temporal abscess is practically never found unexpectedly at autopsy in clinical cases properly observed. The data concerning contralateral and homolateral disturbances of hearing are still equivocal, both on account of the sensorial state of the patient and on account of the frequent involvement of the peripheral auditory apparatus in these cases. Aphasic symptoms are characteristic in left-sided lesions. The sensory aphasia of Wernicke does not ordinarily develop from the temporal lobe abscess as it usually occurs, since the cortex and white matter of the superior temporal gyrus are not usually involved until late. Oppenheim pointed out that the lesion characteristically cuts the association paths rather than the auditory centres themselves, so that an amnesic aphasia supervenes in which the patient speaks little and has lost command of many words. The understanding of terms for simple things remains intact. Slight paraphasia and inability to name objects, with maintenance of power to repeat words, are shown. The abscess may disconnect the auditory and the optic centres and thereby effect a partial word-deafness with maintenance of the understanding of words without visual content. Hemianopsia may be produced by deeper lesions. Contralateral motor phenomena are paralytic rather than epileptic in character, and are probably best regarded as due to involvement of the internal capsule. Muscular rigidity, increased deep reflexes, diminution in the abdominal reflexes, development of the Babinski reflex occur. Macewen and Körner pointed out that oculomotor palsy is frequently found on the side of the lesion. Oculomotor palsy and contralateral facial and brachial monoplegia, with other abscess symptoms, point to temporal localization.

4. *Cerebellar Symptoms.* The most important cannot be precisely spoken of as focal; but cerebellar ataxia, Babinski's cerebellar asynergy, general muscular weakness, occasionally hemiataxia, Babinski's adiadokokinesis, forced movements, peculiar attitudes of head and body, vertigo and vomiting, nystagmus, convulsions, and various effects of compression of lower structures may be found. Pain in the back of the

head and nuchal region, retraction of the head, and stiff neck are more characteristic of cerebellar abscess than of other types.

5. *Occipital Symptoms.* Beyond hemianopsia and possibly word-blindness there are few characteristic symptoms from occipital abscess.

Further localizing points are mentioned under the diagnosis of abscesses classified below as of various origin: (1) Traumatic. (2) Otitic. (3) Metastatic. (4) Rhinogenic. (5) The so-called idiopathic brain abscess.

**Diagnosis.—Traumatic Brain Abscess.**—Traumatic brain abscesses form about one-quarter of reported cases (Church and Peterson). They are likely to be superficial and situated directly below the locus of trauma, and naturally affect the convexity, particularly the frontal and parietal regions, more often than other parts of the brain. In contradistinction to localized traumatic leptomeningitis, which may develop very shortly after the trauma, traumatic brain abscess begins to show symptoms only after several days (*acute traumatic brain abscess*) or after a long latent period of weeks to years (*latent traumatic brain abscess*).

In the *acute traumatic form*, the interval between injury and development of frank *symptoms of abscess* may be occupied by signs of brain laceration or by certain meningeal symptoms, followed by a somewhat sudden development (several days to two weeks after trauma) of the signs of heightened brain pressure with fever. Whereupon, with the development of focal signs, the picture of abscess becomes complete. Involvement of motor parts of the brain is obviously more often the rule in traumatic than in other forms of brain abscess, so that both the history of trauma, with local appearances, and the course of the disease, with its focal signs, somewhat simplify the diagnosis. Nevertheless, it may not be easy to tell such cases from cases of suppurative meningitis. It is said that muscular hypertonus and generalized hyperesthesia are less apt to develop in abscess cases. The cortical epilepsy or monoplegic symptoms, which are not infrequent in encephalitis or with cortical cysts of softening, sometimes occur in traumatic brain abscess, but speedily develop into hemiparesis or hemiplegia, possibly with aphasia. Sometimes the hemiplegia comes on suddenly. Again, there are rather characteristic short remissions which indicate that the process of abscess development is by no means so free as that of a meningitis of similar localization. Oppenheim states that the focal signs of brain abscess are less likely to be masked by general signs than are the focal signs of meningitis, an observation which probably signifies that abscess may proceed for many hours or days without interfering with intracranial pressure. The fever which may characterize the initial onset of pressure symptoms need not be a prominent feature in the later course. In fact, absence of fever as well as slow pulse are commonly thought to favor the diagnosis of abscess when the symptoms of the disease are fully developed. The frequent co-existence of abscess and localized meningitis, as emphasized by Huguenin, must not be forgotten. In the absence of surgical interference, death may ensue within three weeks.

In the *latent traumatic form* we find a similar preference for frontal, central, or parietal localization, but, although these abscesses are certainly related to overlying local trauma, yet they are not due, in the opinion

of most writers, to direct infection from superficial injured tissues. Possibly subinfection with organisms, circulating in the blood but derived from some quite different source, may account for the lighting up of an acute inflammation in the mechanically injured brain tissue. These foci would otherwise, and doubtless often do, gradually heal and in the end produce quite imperceptible functional changes. A careful inspection of the latent periods in such cases may often reveal suggestive symptoms, as a rule, of an irritative or "functional" character. The frank onset of the disease is attended with headache, perhaps localized and accompanied by pain on percussion of the adjacent bone. Vomiting, vertigo, tremors, slow pulse, are characteristic, and a large number of cases fail to show fever, but may even exhibit a subnormal temperature. Such abscesses are consistent with but slight alterations of consciousness, but investigation usually discovers some change, if nothing more than unusual somnolence; such changes when observed are often variable. Changes in the eye-grounds are not constant. Emaciation and gastro-intestinal disorders are frequent. There is great possibility of confusing such latent abscesses with brain tumor; in the differential diagnosis we are aided by the occasional febrile attacks, the rapid progression, and the lack or late development of eye-ground changes in abscess.

An instance of blood-borne infection of intracranial abscess with *Bacillus typhosus* in a traumatic case under treatment for typhoid fever has been reported by Gurd and Nelles.<sup>1</sup>

**Otitic Brain Abscess.**—One-third to one-half of all cases of brain abscess are said to follow purulent otitis media. Of course, only a small proportion of such otitides give rise to brain abscess, and then often many years after the onset. Mastoiditis, temporal bone disease, and the so-called cholesteatoma (of the otologists), and cases of otitis media which do not secure free drainage (either naturally or surgically) are especially dangerous. Less frequently brain abscess follows immediately upon an acute otitis media. A somewhat rare complication is brain abscess ensuing upon the temporal bone disease found in diabetes. Otitis abscesses of the brain are rare in cases which have never undergone perforation.

Like the traumatic forms, otitic brain abscess is usually solitary. Its characteristic locus is the temporal lobe, and if in the right temporal lobe it may yield no localizing sign, but if in the left, may yield word deafness or sensory aphasia, permitting accurate local diagnosis. Naturally, the general symptoms may hinder the detection of this feature. Let such an abscess spread inward, and a spastic hemiparesis, possibly associated with hemianesthesia and hemianopsia, will ensue, as a result of pressure upon, or destruction of, the underlying white paths. Oculomotor and abducent disorders, especially ptosis, may be associated with the other symptoms through pressure upon the corresponding nerves.

*Temporal lobe abscess* is about twice as frequent as cerebellar abscess following ear disease (Heimann,<sup>2</sup> 428 temporal, 198 cerebellar; Neumann,<sup>3</sup> 336 temporal, 196 cerebellar). Cerebellar otitic abscesses in about nine cases out of ten (Neumann) produce symptoms. One group of symptoms

<sup>1</sup> *Annals of Surgery*, 1908, xlvii, 4 to 9.

<sup>2</sup> *Arch. f. Ohrenh.*, 1905, lxvi, 67.

<sup>3</sup> *Der otitische Kleinhirnsabszess*, 1907.

(*vestibular nystagmus*, *vertigo*, and *vestibular ataxia*) is dependent upon involvement of Deiters' nucleus. Inasmuch as labyrinthitis and cerebellar abscess often co-exist, the diagnosis of vestibular nystagmus may be difficult. The ataxia of cerebellar otitic abscess is, according to Barany, a characteristic compound of vestibular and cerebellar features. Peculiar correlations between the direction of nystagmus, the position of the head and trunk, and distribution of ataxia have been described. Nystagmus to the diseased side, described as Neumann's sign, has been confirmed in three of five cases of cerebellar abscess by Hegener. Another group of symptoms (hemiparesis and hemiataxia) depends upon interruption of white paths in the cerebellar tissue. Rapid emaciation and anemia have characterized many cases (Okada). Uncomplicated cases are apt to run an afebrile course. A slow pulse, without correspondence with such fever as may occur, as in other processes, shows heightened intracranial pressure. Occipital pain is *not* especially characteristic of cerebellar abscess; on the contrary, frontal pain is not infrequent (spread of irritation from a recurrent branch of the ophthalmic division of the trigeminal nerve, supplying the tentorium, to other branches of that division, Krause).

Temporal lobe abscess and cerebellar abscess are often difficult to distinguish. Hemianopsia is uncommon in cerebellar abscess. The speech disturbances of cerebellar abscess are rather bulbar than cerebral in character. The hemiparesis of cerebellar abscess is homolateral; of temporal abscess, contralateral. Palsies of isolated eye muscles are the rule in cerebellar abscess, whereas ptosis, external strabismus, and oculomotor palsy characterize the temporal cases. The pain of temporal lobe abscess cases is temporal or parietal, as against the occipital or frontal pain of cerebellar abscess. Nystagmus, stiff neck, and sensitiveness of neck tissues are characteristic of cerebellar cases.

The law of Toynbee, according to which disease of the tympanic cavity gave rise to cerebral abscess, whereas disease of the external meatus threatened rather the lateral sinus and the cerebellum, and labyrinthine disease was likely to lead to bulbar involvement, has been much modified by more recent knowledge as to possible paths of infection. Huguenin pointed out that the cerebellum could be involved directly along the facial and auditory nerves.

Cases of Stokes, of Habermann, and of Bloch and Hechinger indicate that the sense of smell should be tested in this group of cases. Twice the anosmia was homolateral with abscess, once contralateral. Dench<sup>1</sup> published an analysis of 102 recorded cases of cerebellar abscess (33 recoveries) and 100 cases of cerebral abscess (52 recoveries) of otitic origin.

**Metastatic Brain Abscess.**—The diagnosis of metastatic brain abscess (about 10 per cent. of all cases, Gowers) must take into account the predominant sources for embolism. Bronchiectatic abscesses, pulmonary gangrene, lung abscess, pleural empyema, ulcerative endocarditis, pyemic processes of very various origin, often permit the production of brain

<sup>1</sup> Otitic Brain Abscess, *Amer. Jour. Med. Sc.*, 1907, cxxxiv, p. 692.



abscesses, and, as in other hematogenous brain diseases, the territory of the Sylvian arteries is especially prone to involvement. It is also stated that the left Sylvian region is the locus of election for metastatic abscess. Metastatic brain abscess is more often multiple than single and the symptoms more closely resemble those of the acute traumatic form than those of otitic abscesses. Embolic in origin, this disease is apt to have an apoplectiform onset, but the effects of the septic emboli are likely to continue and create sudden fresh attacks, due either to extension of local processes or to the accession of more emboli. The emboli are usually small and carried to the small branches supplying the cortex cerebri or its subjacent white matter.

The Sylvian predilection of such emboli secures a predominantly motor set of symptoms for these cases. But in some instances the foci are so many that localization is impossible. Eye-ground changes usually fail to develop in the acute course of these cases, nor are vomiting and vertigo such prominent symptoms as in most other forms of brain abscess.

**Rhinogenic Brain Abscess.**—Gerber<sup>1</sup> collected from the literature 66 cases of brain abscess following frontal sinus disease. In the majority of cases the posterior wall of the sinus was diseased, and the abscesses are usually found involving the gyri of the orbital surface, and vary in size up to that of an orange. The characteristic absence of focal symptoms, the occurrence of frontal sinus symptoms, and the complicating presence of meningitis symptoms render the diagnosis difficult. Eye-ground changes are not uncommon, so that a diagnosis as against brain tumor has to be guarded. If, after frontal sinus operation, the headaches continue or increase, and fever, loss of appetite, alteration of general attitude, slowness of speech, restlessness alternating with apathy set in, orbital brain abscess may be suspected (Gerber). Later, several of the general symptoms of brain abscess may set in.

**Idiopathic Brain Abscess.**—Huguenin was forced to the opinion that idiopathic suppuration of the brain does not exist. Gowers states that in about one-sixth of the total number of cases no cause for the abscess can be discovered. He believes that forgotten trauma is an important feature of some of these cases, but regards the whole matter as an open question. Oppenheim also suggests forgotten trauma, and thinks that infectious diseases may at times produce suppurative disease in the brain without foci elsewhere (possibly through an otitis?).

**Course and General Diagnostic Features.**—The symptoms of brain abscess may all occur separately with meningitis, sinus thrombosis, and brain tumor, especially if these be complicated by extension of purulent or destructive processes. Moreover, syphilis sometimes yields a similar picture, and the difficult differentiation of symptoms of labyrinthine and central origin may often leave the nature of a case in doubt. In the absence of ear disease, disease of the nose and its communicating sinuses, bronchiectasis, empyema, or other peripheral source of metastasis, the chances are against the diagnosis of brain abscess. The history of trauma is unfortunately often dubious of interpretation.

<sup>1</sup> *Beitr. zur Anat. Path. und Klinik der Stirnhöhlen*, 1909.

As against *meningitis*, abscess more often produces hemiplegia, monoplegia, or aphasia, whereas irritative symptoms are more common in meningitis. Cerebellar abscess, however, may produce the same retraction of the head and stiffness of the neck as meningitis. The absence of fever is against meningitis; its presence favors neither diagnosis.

As against *sinus thrombosis*, the lack of high fever, of rapid pulse, and of tenderness and swelling over the internal jugular vein at its origin may be decisive for abscess. The occasional co-existence of the two lesions must be remembered. As against *brain tumor*, double optic neuritis is less frequent in abscess. Bramwell states that he never commits himself to a positive diagnosis of intracranial tumor unless suppurative ear and nose disease can be excluded.

In the majority of cases three or four stages may be distinguished: (1) The initial stage (lasting a day to a week), characterized by headache, vomiting, slow cerebration, confusion, delirium, fever, and rapid or slow pulse, with possibly stiffness of the neck, pupillary differences, convulsive phenomena, and the relative *absence* of focal symptoms. (2) The latent period (lasting weeks, months, or years, on the average one to three months), either absolute, when the symptoms are completely lacking, or relative, characterized perhaps by episodes of headache, vomiting, vertigo, and convulsions, and failing to show any constant phenomena save, possibly, depression. (3) The stage of abscess symptoms properly speaking (lasting some days or weeks), characterized by the signs of heightened intracranial pressure and the extension of suppuration. (4) The terminal stage.

**Prognosis and Treatment.**—Left to itself, brain abscess kills, despite the extraordinary periods of latency often reported. It may kill through heightening of intracranial pressure, perhaps effected by spreading of suppuration into the ventricles or the meninges. The treatment is essentially surgical. Oppenheim counsels opening the skull and searching for suppuration in all cases in which the diagnosis of traumatic brain abscess is certain or even probable. The indications for operation in otitic cases are scarcely less decisive. Investigation of the cerebral and cerebellar conditions in the course of otological operations must be left to the judgment of the surgeon. Every large clinic has cases in which an abscess cavity in these regions has been "just missed" by the operator. Oppenheim goes so far as to state that purulent pachymeningitis, sinus thrombosis, or even beginning pyemia do not contra-indicate operation. The onset of coma does not prove that surgery would fail. Obviously operations upon metastatic abscesses are less likely to prolong life, and multiple pyemic abscesses are beyond our present treatment. But experience shows that metastatic abscesses are not seldom single and warrant operation upon diagnosis (Krause).

The mechanical increase of intracranial pressure either directly or through the effects of drugs or foodstuffs upon the vascular system should be avoided. Therapeutic measures against headache (narcotics, morphine, ice bags) and attention to the possibility of bedsores and the condition of the bladder and rectum, to say nothing of general nursing and hygienic measures, are obvious indications.

## CHAPTER XII.

### DISEASES OF THE CEREBRAL BLOODVESSELS.

By HENRY M. THOMAS, M.D.

**Anatomy.**—The blood reaches the cranial cavity by four arteries, two on each side, and these also connect with the rich plexus about the spinal cord, which in its turn receives blood at various levels by arteries which enter the spinal column along with certain of the nerve roots. There are then three arterial systems supplying the brain—the internal carotids, the vertebrals, and the spinal plexus. The relative importance of these varies a great deal, especially that of the spinal plexus, which is of comparatively slight importance in man.

The *vertebrals* which arise from the subclavian arteries pass through the canals formed by the foramina in the transverse processes of the upper six cervical vertebræ, and pierce the dura mater between the atlas and the base of the skull. After entering the subdural space they converge along the lateral and anterior aspects of the medulla oblongata to the median line, where near the inferior border of the pons they unite and form the basilar artery. Each vertebral, in its intradural course, gives off a varying number of branches, which pass over the lateral to the dorsal aspect of the medulla. Any one of these branches may be much larger than its fellows and form the posterior inferior cerebellar artery which reaches the inferior surface of the cerebellar hemisphere, over which it ramifies. The lower vertebral branches are connected intimately with the spinal arterial plexus, and more or less with the branches above. The posterior spinal arteries rise from this network of arteries formed from the branches of the vertebral, or directly from one branch, as the posterior inferior cerebellar artery. Shortly before the vertebral arteries join to form the basilar artery each gives off the relatively large twig which passes over the ventral aspect of the medulla to join and form the unpaired anterior spinal artery.

The *basilar*, in its course over the pons, gives off numerous small branches, and from it also arise the anterior inferior, or middle, and superior cerebellar arteries. When the basilar reaches the superior border of the pons it divides in a T-shaped manner, and may be said to end in the two posterior cerebral arteries, which, passing above the tentorium, apply themselves to the inferior and posterior aspects of the cerebral hemispheres.

It is evident that the vertebral arteries entering the posterior fossa of the cranium supply all the important structures lying in this fossa. This vertebral system anastomoses freely below with the arterial system of the spinal cord, and ends above in the posterior cerebral arteries, which form the posterior pair of the three great arteries of the cerebral hemispheres.

From each of the posterior cerebral arteries, soon after its origin, is given off a vessel, the posterior communicating artery, which unites the vertebral system with that of the internal carotids and helps in the formation of the circle of Willis.

The internal carotids enter the cranial cavity through the carotid foramen. Each pierces the dura mater and is continued directly into the middle cerebral artery, which is connected with the posterior cerebral artery by the posterior communicating artery. It also gives off the anterior cerebral artery, which passes forward and toward the middle line to be supplied to the medial surface of the cerebral hemisphere. Just in front of the optic chiasm the two anterior cerebral arteries are united by a short trunk, the anterior communicating artery, and thus the circle of Willis is completed.

There is the freest communication between the large arterial trunks at the base of the brain, and the circulation of the two sides is intimately connected. It must be borne in mind that there is considerable variation in the size of the communicating branches, and that the circle of Willis is not so regular as the diagrams would indicate.

The blood for all parts of the brain comes from a common arterial plexus or reservoir, situated in the middle line at the base of the brain. The vessels which nourish the structures at the base of the cerebral hemispheres, the basilar nuclei, etc., are given off directly from this plexus, or from large arterial trunks shortly after they leave it. These nutrient basal arteries penetrate the brain to a greater or less extent, and appear to be end arteries. The cerebral cortex gets its blood by the anterior, middle, and posterior cerebral arteries. Each of these supplies a more or less definite area, but these are not absolutely distinct, as they overlap and there are actual anastomoses by arterial branches of fair size (from 0.5 to 1 mm.) between the vessels of neighboring areas.

The nutrient vessels of the cortex, so-called cortical vessels, are not given off from the larger arterial twigs, but from the smallest vessels of the pia mater, which have arisen by division of the larger branches. In the rich arterial mesh of the pia one is able to find occasional anastomoses between arterial twigs of fair size, but there does not appear to be a true pial plexus in the sense of Huebner. The nutrient cortical vessels enter the cortex at right angles and descend to a greater or less depth. They give off branches to the cortex and the white matter beneath it, and seem to be practically end arteries, although anastomoses between the branches of neighboring vessels have been described.

The vessels of the cortical system which pass to the white matter almost reach to the basal nuclei. They, however, make no anastomoses with the system of basal nutrient arteries, and the two systems seem quite independent. Beever's<sup>1</sup> injections show as never before the area supplied by the different cerebral arteries, and his work settled many disputed points. The drawings of the arterial supply of the basal nuclei by Aitken<sup>2</sup> are also most instructive.

<sup>1</sup> *Phil. Tr. Lond.*, 1908, cc, 1; *Brain*, 1907, xxx, 1.

<sup>2</sup> *Boston Med. and Surg. Jour.*, 1909, clx.

The blood, after having passed through the capillaries, is collected by the small veins and taken to the surface of the brain, where they join in the formation of the venous plexus, which empties its blood by the larger veins into the venous sinuses. The veins on the surface of the brain are less numerous than the arteries, but are of larger size and have a more superficial course. The various sinuses are large venous reservoirs, lying in folds of the dura mater. Most of them are protected on one side by the skull itself. They are intimately connected among themselves and all empty into the internal jugular veins. The intracranial venous plexus also connects with the veins of the rest of the body by other ways. Chief among these is the anastomosis between the cavernous sinus and the veins of the orbit, which, in their turn, connect with the superficial veins of the face. The other less important connections need not be mentioned, except to point out that there are numerous anastomoses between the veins which run in the diploë and the intracranial system on one side and the veins on the outer surface of the skull on the other.

**Physiology.**—Within the skull we have the brain with its membranes, blood, and cerebrospinal fluid. There is much dispute about the exact conditions which regulate the circulation of the brain. We may regard the skull as a closed box, protecting its contents from the direct action of atmospheric pressure. The total amount of blood within the skull at any instant varies but little from that at any other. This is true because the brain substance itself is incompressible, and the small amount of cerebrospinal fluid is the only thing that can vary. Although the total amount of blood varies but little, the proportion between the arterial and venous blood may and does vary greatly.

The arteries of the brain have well-developed muscular walls, and contain numerous nerve fibres, and it seems certain that they do at times change their caliber independently of the general arterial pressure. But it is generally stated by physiologists that there is no definite proof that the cerebral vessels are under effective vasomotor control, and the view of Leonard Hill, that the cerebral circulation passively follows changes in the general circulation, is widely accepted. Most experimenters failed to get any evidence of direct vasomotor action until Müller and Siebeck<sup>1</sup> reported experiments in which they cut and stimulated the cervical sympathetic, and obtained evidences of definite vascular changes in the brain; and Wiggers<sup>2</sup> has shown that the cerebral vessels, as well as the renal vessels, react to various drugs in very characteristic ways, which he believes is due, at least in a large part, to the effect of the drugs on the nerve endings. It takes a much stronger solution of adrenalin, for instance, to cause the arteries of the brain to contract, than is effective on the vessels of the kidney. Surgeons have noticed the pial vessels change their caliber under the influence of drugs (Cushing).

Therefore, it is probable that the cerebral vessels are under a more or less effective vasomotor control, but much must yet be done to establish

<sup>1</sup> *Zeitsch. f. Exper. Path. u. Therap.*, 1907, iv, 57.

<sup>2</sup> *Amer. Jour. Phys.*, 1907, xx.

this on a satisfactory basis, and in any case the cerebral circulation as a whole does follow passively that of the general circulation. When the blood-pressure is raised, the blood enters the skull with increased force, and a greater quantity passes through the brain in a given time.

Consciousness depends upon a sufficient amount of blood reaching the brain, particularly the cortex, and life itself depends upon arterial blood passing through the medullary centres. To the vasomotor centre is delegated the function of maintaining an adequate blood supply to the other vital centres and to the brain in general. When there is a lack of arterial blood reaching this centre, it is thrown into strong activity and causes a contraction of the arterioles in the splanchnic and skin areas; the general blood pressure is raised, and the blood enters the brain with augmented force and floods the capillaries with arterial blood.

The intracranial pressure, that is, the force which the brain exerts against the skull, depends upon the blood-pressure, and is equal to the pressure of the blood in the venous sinuses, which, under normal circumstances, is the same as the general venous pressure. If the intracranial pressure be raised above the general arterial pressure, no blood can enter the skull, and the animal will die. In Cushing's<sup>1</sup> experiment it was shown that when this pressure became so great as to embarrass the medullary centres there was a corresponding rise of general arterial pressure. If the intracranial pressure was again increased the vasomotor centres answered again with a rise of arterial pressure. This could be repeated until the vasomotor centres were exhausted.

When this regulating mechanism is disturbed, various results follow as, for instance, when the sudden removal of something that has caused continuous pressure on the abdominal vessels, as ascitic fluid, is followed by alarming symptoms. In this case the vasomotor influences controlling the splanchnic area have not been called on for some time, and the centre itself is weakened, so that when a sudden demand is made to compensate for the accustomed external support to the bloodvessels, it is unable to respond, the blood collects in the splanchnic vessels, the patient becomes unconscious and may die.

While under ordinary circumstances the vasomotor mechanism and the tonicity of the muscles of the abdominal walls compensate perfectly for the change from the horizontal to the upright position, *i. e.*, for the effect of gravity upon the column of venous blood from the heart to the feet, in asthenic states, as after severe illness, the compensation may be very imperfect. When such is the case, if the patient stands, or, at times, even if he sits up in bed, his heart beats more rapidly, he becomes giddy, and may faint. The change in the pulse rate with a change in position is a fair indication of the vasomotor control, for the heart endeavors to make up for this incompetence.

Under various conditions the heart itself may become so weak as to be unable to keep the brain properly supplied with arterial blood. The ordinary fainting fit is, at least in part, an example of this. Under the influence of emotion the heart's action becomes weak, the vasomotor

<sup>1</sup> *Mitt. a. d. Grenz. d. Med. u. Chir.*, 1902, ix; *Am. Jour. Med. Sc.*, 1902, cxxiv.

centre is inhibited, and, in consequence, the abdominal bloodvessels become dilated, blood-pressure falls, and the heart is no longer able to drive the blood back to itself against the force of gravity; the blood accumulates in the abdominal veins, the heart empties, cerebral circulation fails, and unconsciousness occurs. The sudden loss of consciousness in epilepsy has also been ascribed to a transient paralysis of the heart (Russell). In Stokes-Adams disease the cerebral symptoms, attacks of unconsciousness, convulsions, and apoplecticiform seizures, are due to cerebral anemia, caused by the temporary cessation of the ventricular systole. The extreme example of this cardiac weakness is paralysis of the heart muscle from failure of the coronary circulation, which is the usual cause of sudden death.

Movements of the chest in respiration have an important effect upon the circulation in general and on that within the skull in particular. With every inspiration the blood is sucked into the heart from the veins, and the descent of the diaphragm, by increasing the pressure on the abdominal veins, tends to force the blood into the heart. During expiration, the intrathoracic pressure is increased, and the entrance of the blood into the heart is impeded. Arterial blood-pressure is influenced but slightly by the respiratory movements. The unconsciousness, or indeed, death, which at times follows forcible compression of the chest, is due to failure of the cerebral circulation caused by the inability of the heart to fill itself with blood, as well as to the damming back of the blood into the venous sinuses.

*Intracranial pressure* has been shown to be equal to the venous blood-pressure within the sinuses and to follow every change in this. The column of venous blood between the brain and the superior vena cava is uninterrupted by competent valves, and every change of pressure in the cava is transmitted directly to the sinuses and veins of the brain. The brain dilates with each pulse beat, but much more with each expiration, that is every rise in venous pressure. Intrathoracic pressure is increased in expiration, and this causes an increase in the pressure of the cava, the jugular, and the brain sinuses. During prolonged expiratory efforts, the venous blood is, as it were, dammed back into the brain, intracranial pressure rises, and, as less arterial blood can pass through the cerebral circulation, the symptoms of anemia may follow, as when a crying child holds his breath until he becomes unconscious.

Intrathoracic pressure makes it more difficult for the heart to fill itself with blood, and affects the circulation also in this way. Strong respiratory efforts against an obstruction may cause very marked changes in intrathoracic pressure. If the glottis be closed and forced expiration be attempted, a normal, negative pressure of the thorax becomes markedly positive, and may far exceed normal pressure in the intrathoracic veins. This is conveyed directly to the cerebral veins and capillaries, and it is not uncommon to have hemorrhages within the brain substance following such an effort. These conditions accompany a strong effort, as straining at stool, lifting a heavy weight, or coughing severely. The importance of preventing, so far as possible, any obstruction to respiration during the course of apoplexy follows directly from what has been said.

The venous outlets from the skull are so large and the anastomoses so free that they must all be obstructed to cause venous congestion of the brain. It is for this reason that thrombosis or ligature of one of the sinuses is not necessarily followed by any symptoms, and, indeed, the superior vena cava itself may be obstructed, as by a tumor, without symptoms. In this last case the blood from the vein reaches the heart through the craniovertebral veins and anastomoses through paths to the inferior vena cava. If all the veins of the neck are compressed as by a tight band or by strong flexion of the neck, the circulation may be impeded, and this is important under pathological conditions.

**Hyperemia.**—The brain is extremely sensitive to changes in the amount of blood which it receives. Many symptoms have been ascribed to anemia and hyperemia of the brain, but there is little agreement as to the symptoms which depend upon these two conditions, and, indeed, upon just what is meant by them. Hyperemia is sometimes divided into passive and active; passive hyperemia meaning the damming back of blood into an organ by impeding the outflow from the veins, in fact, a venous hyperemia and an arterial anemia. The symptoms which follow must be largely due to a lack of arterial blood, and are, therefore, quite similar to those which depend upon anemia.

Active hyperemia of the brain is due to anything that causes an increase of flow through it, *i. e.*, anything that makes the difference between arterial and venous pressure greater; for instance, an increase of the general arterial pressure or a lowering of the venous pressure in the sinuses. Such changes must be occurring all the time, and we know of no symptoms due to them, and it is hard to believe that any harm follows this increase in the supply of arterial blood.

**Anemia.**—This may be general or local, *i. e.*, the brain as a whole may take part with the rest of the body in a decrease of the total amount of blood, and it may be effected by a change in its quality. On the other hand, the circulation of the brain itself may be altered as a result of local conditions, by ligature of one of the vessels that enter the skull, or by disease of any of the intracranial vessels.

The symptoms due to anemia associated with general loss of blood make up most of the clinical picture following hemorrhage, and were the symptoms upon which the older physicians relied when bleeding was a common therapeutic measure. Since this procedure has become so infrequent the interest in the symptoms of hemorrhage has been largely confined to the surgeons, who study them as the chief part of surgical shock. When the loss of blood is sudden and of sufficient quantity, the patient faints. He first experiences a sense of vertigo, may have ringing in the ears, dimness of vision, and a rapidly increasing insensibility. The breathing becomes shallow and intermittent, more or less of the Cheyne-Stokes type; the pulse is feeble and rapid; the extremities are cold and covered by profuse perspiration. At times, especially when the bleeding has been rather slow, the patient may have general convulsions before loss of consciousness occurs, or, indeed, he may die suddenly without any marked premonitory symptoms, having retained consciousness until the end. This accident is more apt to occur when the patient,



who has lost a considerable quantity of blood in a horizontal position, stands or sits up; or even without any change of position, when the hemorrhage has been great enough to seriously embarrass the heart.

When the central nervous system is supplied with blood of a poor character, as in grave anemias, symptoms of malnutrition, more or less pronounced, are apt to follow. Headache, vertigo, tinnitus, and muscular weakness may, at least in a large part, be thus accounted for. On the other hand, it is remarkable how well and how long at times the nervous system performs its functions without apparent difficulty, even when the blood which nourishes it is far below the normal standard. In most of such cases, however, it will be found that the immunity from symptoms is only relative, and that under stress the brain gives evidence of its insufficient blood supply.

Many symptoms are ascribed to anemia of the brain due to spasm of the vessel walls in circumscribed areas. Thus the transient paralyses in arteriosclerosis, etc., are so explained. This depends upon the assumption of an active vasomotor control, and we have seen how little definite proof there is of such an action. Should it be demonstrated, we may learn to recognize a group of phenomena now little understood.

**Cerebral Arteriosclerosis.**—The vessels of the brain may be diseased either alone or in association with the rest of the bloodvessels. The local occurrence of arterial disease has not as yet been thoroughly worked out, but enough has been done to establish the fact that there may be marked degeneration in one system alone, and that the condition of those vessels which can be easily examined, such as those in the arms and legs, is no absolute criterion to that which may be present in some other system, for instance, the brain.

**Etiology.**—So long as our knowledge as to the production of arteriosclerosis remains incomplete, we can not know why, in one case, the vessels of the brain should be first affected, while in another it is those of some other vascular area. Given the general causes, the wear and tear of life, the acute infections, the intoxications, and the conditions which keep up an increased blood-pressure, we may perhaps assume that those organs upon which the greatest amount of stress is brought would be those in which vascular changes would first appear. We might expect, therefore, in individuals whose activities have been largely intellectual to find the cerebral vessels affected more often than in others whose work has been almost entirely physical. In a general way this appears to be true, although there are so many modifying factors that it is of very little practical value. The brain does not function only during so-called intellectual processes, but takes part in most of the bodily activities, and as cerebral circulation follows passively the changes in that of the rest of the body, the cerebral vessels are under constant strain, and almost as much so in the laborer as in the student.

Certain infections, syphilis for instance, appear to have a peculiar affinity for the cerebral vessels. Syphilitic endarteritis is, indeed, usually distinguished from other alterations of the vessel walls, and is apt to be associated with a gummatous involvement of the meninges. Syphilis, with the other infectious diseases, does also seem to predispose to the

ordinary arterial changes, and arteriosclerosis in infants and children is believed to be frequently due to hereditary infection.

*Age.*—In the brain, as in other parts of the body, degeneration of the arteries may occur at almost any time of life. In hereditary syphilis early arteriosclerosis is common, and at times it follows acute infectious diseases. But generally arteriosclerosis is a rarity before the fortieth year, when, from that time on it becomes more common with every succeeding decade, but extensive arterial disease may occur before this period.

*Pathology.*—Anatomically, fairly clear-cut changes in the brain, associated with alterations in the vessels, have been described. These consist in focal areas of necrosis, which may be very small, but are at times large enough to be seen by the unaided eye. These are essentially infarcts due to obliteration and sometimes to rupture of the capillary vessels. They are most frequently seen in the gray matter of the cortex, but may be found scattered anywhere in the brain. Associated with these one finds the ordinary large areas of disease due to obliteration or rupture of one of the larger arteries, which explain the occurrence of focal symptoms. The cortical changes are distinguished from those in general paresis by being focal and unaccompanied by the special histological elements seen in this disease. The changes in senile dementia are much more diffuse and have their own special characteristics.

Marie<sup>1</sup> and his scholar Ferrand<sup>2</sup> pointed out the importance of small areas of softening (*lacunæ*) secondary to arterial sclerotic changes, as the cause of hemiplegia in elderly people. These were often associated with hemorrhage into the brain, at times with softening, but in more than half of the cases they occurred alone. The symptoms which he associates with these changes are very similar to those described under arteriosclerosis.

*Symptoms.*—The anatomical changes in the arteries alter their make-up and render them less elastic, more brittle, and prone to gross alterations, such as the formation of aneurisms. The walls may encroach on the lumen and decrease the capacity. As a result, the organ supplied by the diseased vessels receives its blood through vessels of a smaller capacity and with walls which are changed. Thus the amount of blood reaching the brain is decreased, and the interchange between the blood and the nervous tissue is altered. This is entirely aside from the actual plugging of an artery or the rupture of its walls.

Nature provides lavishly, and it is probable that under normal circumstances much more blood passes through the brain than can be used. Certain it is that in not a few cases the cerebral vessels may be markedly altered and the brain continue to perform its functions so that no defect is obvious. This is due not only to the utilization of the normal surplus of food, but also to the reserve force of the circulatory system itself, which tends to compensate for anything that impedes the flow of blood. On the other hand, there are cases in which, although the vessels are much less altered, we ascribe the pronounced symptoms to a change in the circulatory condition; but we cannot explain why symptoms occur

<sup>1</sup> *Rev. de Méd.*, 1901, xxi, 281.

<sup>2</sup> *Thesis*, Paris, 1902.

in these and not in the other cases. There is no considerable evidence upon which to base conclusions. The anatomical investigation is apt to be limited to the larger arteries at the base of the brain, and the clinical notes are far too often so meagre as to make even this of little use in correlation with the symptoms, and to render superfluous in this relation a minute study of the smaller and nutrient vessels of the brain substance. It may well be that these smaller vessels are at times much altered, when the larger vessels are but little affected, or the reverse may be true, and also the arteries of the different areas of the brain may be unequally diseased.

One would expect that as the brain began to feel the decreased blood supply the symptoms would at first, if noticed at all, be indefinite and transient, and that they would differ with the varying distribution of the vessels most diseased. Until our knowledge is much more complete any statement as to the symptoms of beginning cerebral arteriosclerosis must be indefinite. There is a growing tendency to explain a great variety of symptoms as primarily due to an altered blood supply. The following are the more important symptoms usually so explained.

*Neurasthenia.*—In certain cases in which arteriosclerosis has reached a relatively high degree, and which present definite symptoms referable to cerebral vascular lesions, a history is given of recurrent attacks of neurasthenia dating from a relatively early period. The conclusion that these were themselves evidences of beginning arterial disease is attractive; and when, on the other hand, one finds men of about forty, or older, suffering from the protean symptoms which we class under neurasthenia, and the physical examination reveals well-marked arterial changes, we may be excused if we explain one condition by the other.

One chief characteristic of the neurasthenic is that he is easily tired after any effort, and that even the slightest physical or mental work is followed by distressing symptoms. If the brain is nourished by an inadequate blood supply or receives its blood through vessels whose walls are so altered as to interfere with the free interchange of the blood and the tissues, one would expect that it would show some such evidences of distress under activity. The fact that most cases of neurasthenia can be cured by appropriate measures indicates that the condition of the brain is usually only a part of the general bodily state. Even if there be a beginning cerebral arteriosclerosis, the changes may be so slight as to be entirely compensated when the general level of bodily vigor is raised. Cerebral activities are important factors in this general uplift, and one may perhaps thus understand some of the good results of mental therapeutics, even in cases of neurasthenia which seem to have been due to acute organic changes, as proved by the subsequent course.

However, the practical indication is clear that we must keep in mind the possibility, in our cases of neurasthenia, even in young individuals, that the chief underlying factor may be a beginning arteriosclerosis, and we should not neglect to examine carefully for this condition, and if found we should not overlook it in our treatment, no matter how enthusiastically we regard the perhaps more alluring field of psychical therapeutics.

*Headache.*—This is often classed as one of the most common and most persistent symptoms in cerebral arteriosclerosis, and, indeed, it does occur; but the writer has been struck by the relative infrequency of this symptom, and its complete absence occurs under conditions when it might be expected. Thus, a number of elderly patients, although suffering from undoubted general arteriosclerosis, with marked involvement of the retinal vessels, and in whom the blood-pressure remained persistently high, rarely falling to 200 mm. Hg., have entire freedom from any headache. Walton and Paul, in their study of 100 patients, all the subjects of arteriosclerosis, found headache in only 22 per cent., and in this connection they refer to the well-known fact that individuals who have been subject to headache in their youth, often become immune with advancing years; whereas the opposite would be expected were arteriosclerosis in itself a common cause of headache.

On the other hand, one is at times consulted by patients suffering from intense headache, who give a history of having been entirely free up to late middle life, and it is in these cases that we should carefully examine the vascular condition; but even in them it may be the associated changes that are at fault, such as in the kidneys. When arteriosclerosis has advanced so far as to give focal cerebral symptoms, the onset of this is very generally associated with pain and other distressing sensations referred to the head.

*Vertigo.*—Nearly all patients complain of a sense of insecurity of equilibrium, of dizziness, or of actual vertigo. This may be more or less constant, but is more commonly transient and brought on by exertion or change of position. Some patients when placed in a certain attitude suffer at once from a sharp attack of vertigo, as in two patients who could not be shaved in the usual manner, for the instant the barber tilted back the chair they had a vertiginous attack with the sense of impending loss of consciousness and death. In each of these cases this distressing symptom subsided under hygienic and tonic measures. Subjective auditory sensations, as well as true tinnitus, are occasionally associated with vertigo or may occur alone. The presumption is that there is some involvement of the middle or internal ear, but if such is the fact it escapes demonstration. It may be that some cases in elderly people, which we are in the habit of designating aural vertigo, are really to be put in this category. Vertigo, like headache, is commonly associated with the transient cerebral attacks which are very frequent in certain cases of cerebral arterial disease.

*Apoplectiform Attacks.*—It is not uncommon to have sudden attacks due to more or less closure of some one of the cerebral bloodvessels, or, indeed, to their rupture. The resulting paralysis may be permanent, or it may last only a short time. In the first case we believe that there has been a destruction of nervous tissue, but in the second the loss of function is so transient that we assume it to be due to a passing stoppage of the circulation, from the effects of which the brain recovered. It is these latter cases in which we are at present interested, although they are relatively much less common. These patients suffer from repeated, and in some cases very many, attacks of vertigo and headache, associated

with aphasia, or a more or less extensive paralysis of one side or the other, from which they recover completely. These attacks may come on spontaneously or after exertion. They have been brought into relation with the intermittent loss of function which under stress occurs in other organs, whose bloodvessels are diseased—intermittent claudication, angina pectoris, etc., and the assumption of a transient spasm of the walls of the vessels is given as an explanation. Even the term claudication has by some authors been extended to include these cerebral attacks, but this seems a too free use of the term. That the cerebral arteries during these attacks are in a state of spasm is an inference which receives strong support from observations on the retina in certain cases of transient blindness when the actual constriction and relaxation of the sclerosed central artery is to be noted (Zentmayer).

*Convulsions.*—Convulsions, either general or focal, may be associated with the apoplectiform attacks, but perhaps more frequently the subject of cerebral arteriosclerosis suffers from convulsions which have the usual characteristics of epilepsy. The occurrence of such seizures in individuals who have reached middle life always makes one suspect the presence of this condition. Typical Jacksonian attacks at times recur for years after the occurrence of a cortical lesion.

*Mental Symptoms.*—If the mental states which so frequently accompany neurasthenia are included under this heading—the inability to continue mental effort for any length of time, the forgetfulness, the irritability, the depression of spirits, etc.—we may say that mental changes occur early in cerebral arteriosclerosis. Usually, however, it is not until the disease has progressed much farther that the mental changes are so pronounced as to constitute a real alienation. On the other hand, it has been suggested that in manic-depressive insanity, the varying mental states, with their associated toxic condition, are particularly favorable for the occurrence of arteriosclerosis.

The other types of mental disease which occur late in life, the devolutional psychoses (Farrar<sup>1</sup>), are often associated with vascular changes, and it is difficult at times to determine how much of the symptomatology is actually due to the altered circulatory condition. From these psychoses, occurring during the latter part of life, there has been differentiated a distinct type which is believed to be due to cerebral arteriosclerosis, and the symptoms have been brought into relation with fairly definite anatomical changes. Such patients who may have passed through previous attacks of neurasthenia begin to appreciate that they can no longer accomplish their accustomed mental work, that they tire easily, are unable to undertake new problems, often notice that their memory fails them, that they are unable to recall names, and frequently miss the right word. These symptoms are often associated with attacks of vertigo and other manifestations of arteriosclerosis described above; later definite vascular lesions occur, which, although they may be recovered from, tend to leave the patient on a lower mental level. In other cases these seizures may be the first indication of the process.

<sup>1</sup> *Rev. of Neurol. and Psychiat.*, 1906.

As the mental failure increases the patient becomes unable to carry on his work, is usually apathetic, but his mood may show marked variability, and changes quickly from deep depression to well-being. It is uncommon, however, for this to reach an actual state of exaltation, such as is seen in general paresis. Most patients have a remarkably clear appreciation of their condition, and this is an important help in distinguishing them. The condition may progress to almost complete dementia, but is more likely to be terminated by a vascular accident.

The clinical picture at times closely resembles that of general paresis, and it may also simulate senile dementia. The diagnosis rests largely upon the presence of the more or less characteristic mental conditions, associated with definite symptoms of arteriosclerosis, and the absence of the objective symptoms which point to general paresis, or the typical picture of the senile dement. The age is also of importance as general paresis occurs, as a rule, during the full vigor of maturity, while senile dementia occurs particularly in old age. It is between these periods that the insanities due to arteriosclerosis are apt to develop. The diagnosis may be impossible, and cerebral arteriosclerosis may modify the picture of general paresis and of the psychoses of the devolutional period.

**Diagnosis.**—The cardinal points—(1) thickening of the peripheral vessels; (2) signs of hypertrophy of the left ventricle; (3) heightened blood-pressure; and (4) a slight and variable amount of albumin in the urine—should be kept in mind and looked for when the nervous symptoms suggest cerebral arteriosclerosis. The condition of the retinal vessels is of special significance, for although it cannot be said that alterations in them are always indicative of changes in the cerebral vessels, their condition furnishes the best objective evidences that we have. The ophthalmoscopic picture is quite characteristic.

**Treatment.**—The treatment is that of general arteriosclerosis, but it is essential that the condition should be recognized early, and that treatment should be directed at once to the vascular condition. If the causes are known, measures should be instituted to remove or counteract them as far as possible. Thus, when syphilis is an etiological factor the indication is plain. Usually, it is impossible to determine what has brought about the alteration in the vessel walls; whether the high arterial tension is a cause or effect, and if it be a cause, upon what it depends, and whether the alterations in the kidney are primary or secondary.

**Aneurisms of the Larger Cerebral Arteries.**—True aneurisms of the larger cerebral arteries are relatively uncommon. In Crisp's list, as quoted by Osler (vol. iv, p. 480), among 501 aneurisms, intracranial aneurisms occurred only 7 times. Beadles<sup>1</sup> collected the records of 555 cases of such aneurisms found after death. He classified these in four groups: "(1) Those in which the first indication of a cerebral lesion has been an apoplectic attack due to rupture of the aneurismal sac. (2) Those in which a fatal apoplexy has been preceded by symptoms suggesting a cerebral tumor or other cerebral lesion. (3) Those in which there have been indications of a cerebral tumor only. (4) Those that

<sup>1</sup> *Brain*, 1907, xxx, 285.

gave rise to no symptoms whatever during life, and the aneurism was discovered accidentally after death."

In over one-half of the cases the first symptoms noted were those of apoplexy, and even in those cases in which symptoms did occur these were often trivial, or such as might have been attributed to cerebral arteriosclerosis, such as dizziness, headache, mental weakness, and even insanity. Indeed, not a few cases of cerebral aneurism have been unexpectedly found at autopsy in patients dying in insane hospitals. As aneurismal dilatations depend upon structural changes in the walls of the vessel, arteriosclerosis, or syphilitic disease, similar changes are apt to be present in the walls of the other vessels of the brain, and, as would be expected, not infrequently other lesions are present due to these vascular changes but entirely independent of the aneurism, and it is to these that many of the symptoms may be due.

The general symptoms of brain tumor, violent headache, vomiting, and choked disk, are at times present, but the frequency of their absence is remarkable. Headache particularly seems to be a rare symptom, and choked disk has been only occasionally noted, but Beadles found but few notes of an ophthalmoscopic examination having been made. Aneurisms of the basilar artery give rise more often to focal symptoms than those on any of the other arteries at the base, these occurring in almost one-half the number of cases. This is due to the fact that aneurisms in this situation may implicate the cranial nerves or the important nervous paths which run close to the surface in the pons and médulla. Aneurisms from the posterior communicating, posterior cerebral, and intracranial portion of the internal carotid, and the middle cerebral, have given symptoms with the relative frequency in the order named, but these are almost never so characteristic as to allow of a local diagnosis. It is only in rare instances that the diagnosis of a tumor can be made, and that such a tumor is an aneurism is a deduction that is practically not justified. The classical symptoms supposed to indicate an intracranial aneurism, the subjective noises in the head, and a vascular murmur which could be heard through the skull, are so rare as to be of little value. Beadles was able to find only about "a dozen cases recorded in which some more or less definite sound has been heard in the head of the patient." In none of these was there any record of the head having been examined for an objective murmur, and he was able to find only two cases of "uncomplicated, true, intracranial aneurism in which a murmur had been heard by the examining physician. One of these was an aneurism of the vertebral, and the other an aneurism of the cavernous portion of the internal carotid." The fact that it is so rarely possible to make an accurate diagnosis of an aneurism may perhaps be partially explained by the small size which they usually reach. An aneurism larger than an ordinary marble is rare; one reaching the size of a small hen's egg is a very large one; and Beadles, in speaking of an aneurism, which he so describes, remarks that there have been probably less than a dozen larger. On the other hand, many aneurismal dilatations have ruptured relatively early, when they were no more than small, soft sacs and produced little pressure. It is the slow-growing, thick-walled aneurisms that have

usually given symptoms. In 339 of Beadles' 555 cases, death occurred following a rupture of the aneurism, and it is apparent that their chief importance is in relation to apoplexy.

**Obliteration and Rupture of the Cerebral Vessels.—Thrombosis and Embolism.**—The subject of thrombosis and embolism has been fully considered (vol. iv, p. 526). The reader is also referred to Welch's article in Allbutt's *System of Medicine*, and only points which are of especial interest in the present connection will be referred to here.

**Etiology (Thrombosis).**—The occurrence of thrombi in the vessels of the brain, as in the vessels of the body generally, is conditioned by the state of the vessel walls and by the character and rate of flow of the circulating blood.

The changes in the *vessel walls* are of prime importance, and are believed by many investigators to be essential factors in the production of thrombi. These changes are usually those of arteriosclerosis, and are particularly active when they have affected the intima of the artery, as in the formation of an atheromatous plaque. In arteriosclerosis the walls of the vessels may be thickened, and the lumen so encroached upon, that the formation of even a small thrombus obliterates it. This, however, is more apt to occur in that type of arterial disease associated with syphilis.

Biochemical alterations in the blood may predispose to thrombosis, but whether this occurs in a vessel whose walls are normal is a disputed question, and just what these chemical changes are is not known. In the acute infections of childhood, in typhoid fever, septic infections, etc., thrombosis of the cerebral arteries has occurred. In many of these, however, the thrombi have contained the specific microorganisms of the infection, which also have been found in the vessel walls themselves, so that even here the primary change may have been in the arterial walls. The blood may also carry larger particles about which, when lodged, for instance, at the bifurcation of a vessel, thrombi may develop.

The altered condition of the blood in chlorosis seems to predispose to thrombosis, but the thrombi in this condition usually affect veins, particularly the cerebral sinuses. The change in the rate of flow of the blood is of importance, for when the other conditions favoring the formation of thrombosis are present, a slowing of the blood current may be the determining factor. Thrombi occur especially in those localities where the shape of the channel causes eddies or little whirlpools in the blood current. Such places may depend on the anatomical arrangement, but are more apt to be due to the changes in the vessel walls, for instance, those which produce local dilatations or definite aneurisms. Cerebral thrombosis is generally stated to be more frequent when the circulation as a whole is feeble, due to weakness of the heart or to some other cause.

**Age of Onset.**—The age at which we may look for cerebral thrombosis depends upon the conditions which have been stated. As it at times occurs in association with the acute infections, we may find it in this relation at any period of life, and it is probable that many of the cases of infantile hemiplegia are due to this factor. Thrombosis due to arteriosclerosis is uncommon before the fortieth year, but in later life the conditions favorable to its occurrence are almost constantly present.



Syphilis is a disease usually acquired in early adult life, and the involvement of the cerebral arteries, leading to their occlusion, may occur at almost any time after the primary infection. The writer has seen it occur in the first six months; and it appears that more cases develop in the first than in any other year after the primary lesion, and that they decrease in frequency with every following year. Cerebral thrombosis due to this cause is, therefore, a disease of the prime of life, and most cases occur between twenty and forty.

**Etiology (Embolism).**—Emboli which lodge in the brain are, in the great majority of cases, derived from somewhere between the beginning of the pulmonary veins and the arteries of the cranial cavity. The most common source is the heart itself, as from vegetations on the mitral valves, and they are more frequently associated with mitral constriction than with any other cardiac condition. Emboli are also dislodged from the aortic valves or from thrombi on the arterial walls, and at times, although more rarely, are formed of bits of calcareous matter detached from an atheromatous plaque. If we include microorganisms circulating in the blood as emboli, they may enter the circulation at any point, and pass through the relatively large capillaries of the lungs to be lodged in the capillaries of the brain; but these are not usually so considered.

**Age of Onset.**—There is no particular age at which cerebral emboli are apt to occur. This depends upon the time of life in which the conditions, favorable to their origin, are present. These are for the most part those conditions which produce valvular heart disease, and often occur early in life, but may be present at any time. Embolism is perhaps, on the whole, most frequent between late childhood and middle life.

**Etiology (Cerebral Hemorrhage).**—The rupture of a cerebral vessel depends primarily upon the weakness of its walls, but to some extent upon the pressure of the blood within them. Under normal conditions the vessel walls are competent to withstand any strain put upon them, and it is probable that they must be diseased before they rupture. Various changes may so weaken the wall of a vessel that it gives way, and in rare instances hemorrhages have taken place from arteries affected with syphilitic disease—a process more apt to give rise to thrombosis.

Since Charcot and Bouchard, in 1864, called attention to the presence of miliary aneurisms on the cerebral vessels in cases of apoplexy, it has been generally taught that it was the rupture of these structures that caused the hemorrhage. They are little dilatations, many of which can just be seen with the unaided eye, having a diameter from 0.1 to 1 mm., and very many may be present, scattered along the nutrient vessels of the brain, but particularly along those which nourish the basilar nuclei. There has been much dispute concerning the exact pathological process upon which they depend, which coat of the artery is first diseased and their relationship to arteriosclerosis. Dilatations of various kinds occur on the cerebral vessels, as Virchow showed, and von Monakow<sup>1</sup> points out that it is more or less a matter of choice what dilatations are

<sup>1</sup> *Gehirnpathologie*, second edition, Vienna, 1905, p. 1105. Full literature.

to be considered as miliary aneurisms, and that it is not always possible to distinguish them sharply from other dilatations. Rupture may occur in vessels whose walls have undergone a hyaline degeneration, or those which have undergone alterations secondary to a mechanical injury. All these conditions he considers as exceptions, and concludes: "By far the most common and perhaps the only certain cause for *spontaneous* cerebral hemorrhage must, according to my view, be sought in the bursting of miliary aneurisms. These begin with a degeneration of the muscular coat; an endarteritic process is not essential for their formation."

A. G. Ellis<sup>1</sup> studied by modern histological methods the arteries in 31 cases of spontaneous cerebral hemorrhage, and Pick<sup>2</sup> examined 11 other cases by a special method to determine the occurrence of miliary aneurisms and the source of hemorrhage in each case. These authors conclude that aneurisms of true miliary size are very generally, although not constantly, present in cases of cerebral hemorrhage. Pick was unable to associate them with the hemorrhages. These came from the rupture of large aneurismal swellings, or generally from diseased vessels without such swellings. These aneurisms, the smallest as well as those of larger size, are all, they believe, either dissecting or false aneurisms. The changes in the coats of the vessels, that permit of their occurrence, differ histologically in no respect from those seen in arteriosclerosis, the primary lesions being apparently in the elastic layer of the intima.

All the effective causes in the production of arterial disease may be included among the predisposing causes of cerebral hemorrhage. It is interesting to note how some families show a strong tendency to the occurrence of cerebral hemorrhage at a relatively early period of life. This hereditary weakness of the vascular system may be shown by other vascular accidents—the early occurrence of general arteriosclerosis, angina pectoris, and the frequency of sudden death—even when apoplexy has not been particularly common in the family. Renal disease with hypertrophy of the left ventricle and an increased arterial pressure is frequently present in cases of cerebral hemorrhage. The increased arterial tension is believed to be an important agent in the production of the vascular changes, and is also a factor in the rupture of the vessel. Any great increase of the pressure of the blood within a vessel with weakened walls would tend to cause a break, and so increased arterial pressure is given as a common, exciting cause of cerebral hemorrhage. This increase may be produced during a sudden muscular effort or a fit of anger—conditions which increase the activity of the heart. If the outflow of the blood from the cranial cavity be impeded, as when there is a great rise of pressure on the venous side of the circulation, as during strong expiratory effort, the pressure within the veins throughout the brain is increased, and this increase is extended to the capillaries and even to some extent into the arterioles, and may produce their rupture. This is still more likely to occur when combined with an increase of arterial pressure, as

<sup>1</sup> *Proc. Path. Soc.*, Philadelphia, September, 1909, p. 197.

<sup>2</sup> *Berl. klin. Wchnschr.*, February 21 and 28, 1910.

during strong muscular effort. Such conditions are believed to be present during the intense respiratory spasms of whooping cough, when intense muscular efforts are made, the breath being held, as in lifting heavy weights, etc.—circumstances under which cerebral hemorrhage is said to be common. It will, however, be shown later how infrequently one is able to discover any such exciting cause.

*Age of Onset.*—Cerebral hemorrhage is uncommon before the fortieth year, but may occur at almost any time in life. It is not an infrequent complication of difficult births, but these cases may, for the most part, be considered traumatic, resulting from the pressure on the child's head. True cerebral hemorrhage, however, does seem to occur during the first years of life, and the liability to it increases with each decade.

**Pathology (Cerebral Softening, Encephalomalacia, Thrombosis, Embolism).**—The effect of the occlusion of a cerebral vessel depends upon how completely the blood is shut off from that portion of the brain supplied by the vessel implicated. There is an extremely rich anastomosis between the vessels at the base of the brain, and theoretically the obstruction of a vessel emptying into the circle of Willis should have no effect, but unfortunately the channels which connect the various parts are not always present, or if present are not always of the normal size. Even when the anatomical relations have been quite normal, certain of the connections may be partially or completely obstructed. It is, therefore, impossible to be quite sure of the effect of occlusion, either by disease or ligature of the large vessels. On dogs it has been possible to tie both internal carotids and both vertebrals, provided a sufficient time has intervened between the various ligatures. In these cases the brain receives its arterial blood from the anastomosis with the spinal system. In man, the ligature of one internal carotid is a procedure attended with considerable risk. Serious brain symptoms have occurred in about 25 per cent. of the cases, and death has followed in nearly 10 per cent. (Jordan<sup>1</sup>). Certain of these results may possibly be avoided if the closure of the vessel is done slowly so as to allow the collateral circulation to be established, and it might be possible to ligate safely both common carotids in man if this procedure were followed and sufficient time elapsed between the two ligatures (Leonard Hill), but abnormalities of the circle of Willis must always be taken into account.

The larger arteries after leaving the circle of Willis supply different areas of the brain, but there is more or less free communication between the vessels and it is quite possible to inject the whole brain from any one vascular trunk. The ease with which this is done depends upon the normal condition of the arteries, and in brains of older people, when arterial changes become more common, it may be impossible to do so completely, and the occlusion of one of the larger arteries, at its origin from the circle of Willis, is usually followed by more or less softening. Cases have been recorded in which such occlusion has occurred without any softening having followed. When a thrombus extends, as it commonly does, so as to implicate the origin of the nutrient vessels which

<sup>1</sup> *Verhandl. der Deut. Gesellschaft f. Chir.*, 1907, xxxviii, 83; Becker, *ibid.*, p. 623.

enter the base of the brain, the softening constantly occurs in the gray nuclei which they nourish, for these vessels are end arteries.

The changes which occur in the brain substance, following a closure of its nutrient vessels, have given rise to much discussion. The highly differentiated nervous tissue is probably more susceptible to a decrease in its vascular supply than any other tissue and at times a destruction of this tissue occurs from the blocking of an artery, which is apparently connected with other vessels by free anastomosis, as, for instance, one of the pial vessels. In general, necrosis occurs whenever the vessels which enter the brain substance, the so-called nutrient vessels, are blocked. The area of ischemic necrosis which follows varies in position and size, depending upon the number of vessels occluded.

If the brain be examined shortly after the vascular occlusion has occurred, the necrosed area will be found to have a white or a reddish appearance, being more apt to be red when situated in the cortex where the vascular supply is particularly abundant. There is usually marked œdema which may extend beyond the boundaries of the softened region, and be so great as to cause a definite swelling of a considerable portion of the brain. In certain cases this œdema is the only objective finding to account for the marked symptoms which have followed a vascular occlusion, as in death after ligation of the common carotid.

The microscopic appearance of the necrosed area is characteristic. It is only in its centre that there is a complete disintegration and here one finds a diffuent mass composed of the detritus of the nervous elements, and every now and then a compound granular corpuscle. Around this the tissue shows more or less marked irritative reaction, the blood-vessels are prominent, due particularly to proliferation of the cells of their walls and the collection of leukocytes in the surrounding tissue. Compound granular corpuscles are extremely common; newly formed neuroglial elements are present, and there is a greater or less amount of blood, either in the vessels or free in the tissue; or there may be actual small hemorrhages. The color of the infarct depends upon the number of these red blood cells. The nervous elements in this region are found swollen and in various stages of destruction. If examined after one or two weeks the processes of repair are evident. The red blood pigment is becoming altered, the phagocytes, compound granular corpuscles, leukocytes, etc., have taken up and are removing the destroyed tissue, and there is a distinct increase of the supporting tissue, the neuroglia, and the true connective tissue. If the area is a small one this process may entirely replace it by tissue which usually contains minute cavities scattered through it. In larger areas there is a tendency to cystic formation with more or less definite walls. The process which appears to have the most clinical bearing is the œdema, for it is to this that many of the symptoms may be due. It may be so great as to cause a dangerous increase in the intracranial pressure and account for definite pressure symptoms. To this also may be due many of the initial focal symptoms from which the patient recovers. It is a process which occurs frequently in the nervous tissue, and different theories have been advanced to

account for it. Cannon,<sup>1</sup> in a study of the effects of trauma on the brain, has paid particular attention to this point, and comes to the conclusion that the process depends upon the altered osmotic relations due to changes in the nervous tissue, secondary to vascular disturbances.

When the thrombi which occlude the vessels are infected with pyogenic microorganisms there may be the production of an abscess in the brain.

**Pathological Anatomy of Hemorrhage.**—When a hemorrhage occurs from the rupture of an artery of fair size, blood is thrown out under pressure, nearly equal to that of arterial pressure and much above the cerebral pressure, which is about equal to venous pressure. The surrounding nervous tissue is broken up by the mechanical impact of the blood. The extent of the destruction varies with the amount of blood thrown out and the character of tissue into which it is thrown. The white matter is more easily disintegrated than the gray matter, and a hemorrhage in it is apt to produce a larger area of destruction than when confined to the gray substance. The size of an intracranial hemorrhage varies greatly. It is usually no larger than a walnut, but it may at times occupy almost the whole of the cerebral hemisphere, or be very small.

In the tissue about the actual clot there are usually minute hemorrhages which give it a blood-stained appearance, and a hemorrhage of any considerable size is nearly always surrounded by an area of more or less intense cedema. This cedema may be an important factor in the production of the symptoms, and it is not at all uncommon to find softened areas in the neighborhood of a hemorrhagic focus. If the patient survives the initial shock, processes of repair begin very quickly. There is an endeavor to absorb the clot from the periphery and the formation of a cyst. At first the walls of the cyst are formed of a loose tissue, but this tissue becomes firmer as it gets older, and in cysts of a year or more standing it is a firm connective-tissue capsule, which has on its inner surface a layer of cells of various kinds, stained with the remains of blood pigment. The rapidity of this cyst formation varies greatly, but even in relatively small foci the blood clot can usually be found up to four or five weeks, after which time the cyst formation becomes more and more definite.

### APOPLEXY.

The symptoms following acute vascular lesions of the brain, whether the process be the rupture of a vessel or its occlusion, are in many respects identical; and clinically it is often impossible to determine which process has been effective. For this reason the symptomatology of these different processes is considered together under the general term of apoplexy.

The word *apoplexy* has a most interesting history. It was used by the Greek and Latin medical authors, and originally meant "to strike off or be disabled by a stroke." It appears in middle English, and Chaucer

<sup>1</sup> *Amer. Jour. of Phys.*, 1901, vi, 91.

uses it near the beginning of his "Nonnes Priestes Tale." It was in general literary use in the time of Queen Elizabeth, and Shakespeare makes Falstaff (Henry IV, Second Part, Act I, Scene II) use it as an interesting subject of conversation.

Apoplexy was used in medicine to signify a set of symptoms—a more or less sudden and complete abolition of consciousness with loss of feeling and motion, respiration being maintained. As Sydenham has it: "Profound sleep, utter loss of sense and motion, with the exception of that necessary for respiration." It came to be used almost synonymously with coma, and Abercrombie constantly uses such expressions as "The patient was found in complete apoplexy." The apoplectic attack was often spoken of as a fit or a stroke—the ictus. After it became generally recognized that the cause of apoplexy was commonly a hemorrhage in the substance of the brain, the pathological condition itself began to be called apoplexy, and in the early part of the last century the term was extended to signify any sudden interstitial hemorrhage, as "pulmonary apoplexy," "renal apoplexy," etc. Todd and most of his contemporaries used the term apoplexy as indicating hemorrhage into the brain, but Trousseau insisted on its original symptomatic meaning. At the present time the best English usage is reverting to the original meaning, although the word even yet bears with it a distinct idea of hemorrhage. In the present article apoplexy is used more nearly in its original symptomatic sense, and made to include the cases due to obstruction of the cerebral vessels as well as those following a rupture.

**Occurrence.**—The frequency of apoplexy may be approximately judged from a study of mortality statistics. *The Seventh Annual Report of the Census Bureau* gives the figures for 1906. These cover those States whose mortality reports are deemed sufficiently reliable (a population of 40,996,317, almost one-half the population of the United States). There were 658,105 deaths from all causes, a death rate of 16.1 per thousand. Among these 29,434 were ascribed to apoplexy, and 6933 to paralysis. Many of these latter cases must have been due to previous attacks of apoplexy, although some were probably of an entirely different character. They are, however, considered together, and the total of 36,367 has probably as much chance of being below as above the right figures, for there are many cases included in the mortality statistics under the heading of softening of the brain and a general paralysis of the insane, which should properly be classed among the apoplexies and are not included in these figures. The total of 36,367 deaths from apoplexy and paralysis gives a death rate of 88.7 per 100,000 of the population.

In Baltimore, with a population of 533,699, there were during the year 544 deaths from apoplexy and paralysis. The expected deaths among such a population would be 490. The increased death rate depends upon the number (124) of deaths in colored individuals from these causes (29.5 per cent. of the deaths were in negroes, whereas the negro population forms not quite 19 per cent. of the whole population of Baltimore). This greater liability of the negroes to die from apoplexy and paralysis is quite parallel with their greater liability to diseases of the circulatory system. *The Census Report* shows that among 100,000 of the population

of Baltimore there were 161.3 deaths from such causes among the whites and 327.4 among the blacks.

**Etiology.—Predisposing Causes.**—These are the same as those in the primary vascular lesions. Whatever tends to produce these lesions has a bearing on the production of apoplexy.

**Race.**—The figures from the *Census Report* indicate that there is a distinct tendency in the negro race to diseases of the circulatory system, and also a greater liability to apoplexy. In the writer's series there were 152 negroes among 674 individuals suffering from hemiplegia in whom the race was noted.

**Sex.**—Men are somewhat more liable to apoplexy than women. The proportion among 1000 of the aggregate population within the Registration States was in 1900, 505.8 men and 494.2 women, while among 1000 deaths from apoplexy and paralysis in 1906 there were 504.5 men and 495.5 women. The tables (1913) compiled by the association of the life insurance medical directors show a slightly greater liability of the male policy holders to apoplexy. It is the most common cause of death in men who have taken their policies out after forty-five years of age, while in women of a corresponding age it stands fourth. In the records of the Johns Hopkins Hospital there is a marked preponderance of males, there being 489 males and 251 females, but in this hospital many more men are treated than women.

**Build.**—The popular belief that stout individuals, with short, thick necks, are more liable to apoplexy is confirmed by the insurance tables, for the ratio of deaths due to apoplexy among 10,000 lives exposed to risk is nearly four times as great in the overweight as in the underweight group and about twice that for the standard lives.

**Age.**—The vascular changes upon which the apoplexy depend are in a large proportion of cases conditioned by incidents of the life of the individual, and increase in frequency with each succeeding decade, and the frequency of apoplexy also increases with each decade.

In the mortality statistics a large number of apoplexies are recorded as occurring under one year of age. These must have been composed largely of traumatic apoplexies occurring at the time of birth, and give an undue preponderance to this time of life. The following tables, condensed from the *Census Report*, show the occurrence of apoplexy in the various decades, and give a fair idea of the relative frequency of apoplexy at the different periods. The frequency is, of course, not the same thing as the liability, for the number of cases which occur at any period of life must depend upon the number of people alive at that age. The estimated population in the Registration States in 1907 was distributed among the decades according to the ratios found in the census of 1900, and compared with the number of deaths in each decade which were reported during 1907. The error introduced cannot be great. The relative liability in the different decades is expressed in proportion, taking the liability in the second decade as 1.

TABLE I.—*Deaths from Apoplexy and Paralysis. Census Report for 1907.*

Decade.	Cases.	Proportion per 1000 of deaths.
First . . . . .	670	20.1
Second . . . . .	121	3.6
Third . . . . .	370	11.1
Fourth . . . . .	998	30.0
Fifth . . . . .	2,250	67.7
Sixth . . . . .	4,836	145.4
Seventh . . . . .	8,491	255.3
Eighth . . . . .	9,877	297.0
Ninth . . . . .	5,100	153.4
Tenth . . . . .	544	16.4
Total . . . . .	33,257	1000.0

TABLE II.—*Table Condensed from the Census Report for 1906.*

Population of Registration States in 1907, distributed according to ratios found in 1900.		Deaths from apo- plexy and par- alysis in 1907.	Liability per 100,000 population.	Relative liability.
First decade . .	6,984,883	670	9.6	5
Second decade . .	6,204,278	121	2.0	1
Third decade . .	6,260,922	370	5.9	3
Fourth decade . .	5,171,628	998	19.3	10
Fifth decade . .	3,760,575	2,250	59.8	30
Sixth decade . .	2,550,957	4,836	189.6	95
Seventh decade . .	1,579,307	8,491	537.6	269
Eighth decade . .	734,016	9,877	1,345.6	673
Ninth decade . .	180,990	5,100	2,817.8	1,409
Tenth decade. . .	13,406	544	4,057.9	2,029

The cases in each decade differ somewhat as to their etiology, and each decade will be considered separately. In the Johns Hopkins Hospital there have been 292 cases of apoplexy, *i. e.*, patients suffering either from the acute condition or the subsequent paralysis, in the medical wards among 23,048 medical admissions. In the Out-patient Department 448 cases of hemiplegia and other results of acute lesions were seen. The cases registered in both hospital and dispensary were included only among the hospital cases. The following table shows their occurrence in the various decades:

TABLE III.

Decade.	Males.	Females.	Total.
First . . . . .	58	77	135
Second . . . . .	16	9	25
Third . . . . .	43	19	62
Fourth . . . . .	73	29	102
Fifth . . . . .	88	49	137
Sixth . . . . .	99	44	143
Seventh . . . . .	86	19	105
Eighth . . . . .	20	5	25
Ninth . . . . .	6	0	6
Total . . . . .	489	251	740



**Decade I.**—The cases of hemiplegia in the first decade are of especial interest, and in large part are the result of processes peculiar to that period. In the *Census Report* for 1906 a large proportion of cases of death from apoplexy and paralysis in the first decade occurred in the first year, 621 among a total of 961. These must include those dying shortly after birth with symptoms indicating injury to the brain. In the Johns Hopkins Hospital records there were 135 cases of hemiplegia which had appeared in the first decade. Of these, 32 are classed as congenital, and in 16 the onset occurred during the first year. This can be only relatively accurate, as it is often impossible to determine from the history whether the paralysis was due to conditions present at birth or to those which occurred later. When the parents believed the paralysis to have been always present, and when nothing in the history indicated a later onset, it has been classed as congenital. In other cases the history is so clear that little doubt can be possible.

*Congenital hemiplegias* are in most instances due to accidents occurring at birth. As the head of the infant passes through the pelvic canal of the mother it is subjected to great pressure, which in the vast majority of cases the head is well adapted to withstand, but at times the pressure is great enough to injure the brain, either directly or, as more often, by means of a hemorrhage within the skull. Very rarely the paralysis is the result of a prenatal process, *i. e.*, something that has affected the brain of the fetus before labor has begun. Prenatal pathology in so far as it relates to the central nervous system is an obscure subject. Bal-lantyne,<sup>1</sup> in his large work, has but little to say about it. Gross defects of development in the brain and spinal cord have, indeed, been frequently described. These may be so great as to lead to the entire absence of the whole central nervous system, or of the brain or of a large part of it.

The brains of newborn infants show at times relatively small defects, even when they may be otherwise normally developed. These consist of cavity formation involving to a greater or less extent the cortex and the white matter beneath it (porencephaly), atrophy of the convolutions (microgyria), and sclerosis in certain portions of the cortex or tracts within the central nervous system, the pyramidal tracts especially. Quite similar conditions are frequently found at autopsies performed on cases of infantile cerebral palsies, both in those which are presumably congenital and also in those which appeared during the first years of life. It is to the study of these end results of various initial lesions that our knowledge of such conditions is mostly due.

The term *porencephaly* has been used to describe a great number of conditions. It was first introduced by Heschl, in 1859, but it was not until 1880 that especial attention was drawn to it by Kundrat, from whose monograph our knowledge may be said to date. Kundrat and most other writers believe that these cavity formations are usually due to some prenatal condition, especially to some disturbance in the circulation. These defects occur most often in the central convolutions, and the blood-vessels supplying this region have often, but not always, been found

<sup>1</sup> *Antenatal Pathology and Hygiene, The Fetus*, Edinburgh, 1902, p. 388.

diseased. The vascular obliteration has been ascribed to thrombosis and embolism during fetal life, but there do not seem to be many facts to substantiate this. Inflammation of the brain substance, with secondary softening, is also given as the starting point of the process. That this is actually a cause in prenatal cases seems also to be based on very few facts. Seitz<sup>1</sup> after an analysis of the cases adduced in proof of this belief, first advanced by Virchow<sup>2</sup> in 1867, came to the conclusion that the lesions described under the heading of congenital encephalitis have resulted from cerebral hemorrhage, or are extra-uterine from specific infections, possibly also intoxications. He concludes that congenital encephalitis in the sense that Virchow used it has not been proved to exist. On the other hand, he believes that porencephaly found at birth may be due to a cerebral hemorrhage during fetal life and records the case of a baby who died five hours after an easy birth. At the autopsy a large defect in the anterior and middle parts of the cerebrum was found. The white matter was replaced by a cavity which involved also the cortical substance. The basal ganglia, the pons, cerebellum, and medulla oblongata, were all normally formed. Microscopic examination of the walls of the cavity gave evidence of a former hemorrhage. The mother of the child had received a severe trauma to the abdomen when four months pregnant, which Seitz believed caused an intracerebral hemorrhage in the fetus.

The occurrence of hemorrhage within the fetal brain is certainly rare. Seitz was unable to find in the literature any case quite similar to his, and Ballantyne, in proof that such a condition may occur, refers to only one case, that recorded by Osler.<sup>3</sup> It was in the fetus of a woman six months pregnant, who died of typhoid fever. The hemorrhage was in the left cerebral hemisphere and had broken through into the ventricle. There was also general œdema of the brain.

Gibb<sup>4</sup> records a case of a stillborn child who had a contracture of the left arm and leg and in whose right cerebral hemisphere an old clot was found above the lateral ventricle. The mother of the child had received a severe blow on the right side of the abdomen three months before labor. These cases show that intracerebral hemorrhage does occur during fetal life, both following a trauma to the abdomen of the mother and during the course of an acute illness. Freud refers to a clinical case recorded by Gaudard, in which the mother received a severe blow on the abdomen in the sixth month of pregnancy. The child was born with paralysis of the right arm.

The intra-uterine causes of microgyria and lobar sclerosis are not well understood, but these conditions are generally believed to follow some disturbance of the blood supply to the developing brain. Lack of development of the pyramidal tracts, which is included among the prenatal causes of the cerebral palsies, appears to be the result of the cerebral defect, and not an independent condition.

As has been stated, the great majority of *congenital cerebral palsies* are due to accidents during birth. The association of difficult labor,

<sup>1</sup> *Arch. f. Gyn.*, 1907, lxxxiii, 701.

<sup>3</sup> *Teratologie*, 1895, ii, 13.

<sup>2</sup> *Virchows Archiv*, 1867, xxxviii, 127.

<sup>4</sup> *Lancet*, 1858, ii.

asphyxia, and premature birth with the spastic paralysis of children was pointed out by Little,<sup>1</sup> but that these paralysees were due in most cases to hemorrhage within the cranium was first clearly shown by Sarah J. McNutt.<sup>2</sup> Little was well aware of the frequency of intracranial hemorrhage at the time of birth, but, as at that time the cortex was not known to have anything to do with motion, he believed that the spastic paralysis was due to hemorrhage about the spinal cord. McNutt showed that the hemorrhages incident to birth were apt to be most intense about the fissures of Rolando, and she assumed that the atrophy of the central convolutions was due to a bilateral hemorrhage limited for the most part to this region.

In most instances these cases are associated with abnormal labor, but large and even fatal intracranial hemorrhages do occur when the birth is apparently normal and easy. Marked changes of pressure in the circulation of the infant must take place during labor, especially when placental circulation ceases and that through the lungs is established. When the child is asphyxiated the circulatory conditions present are particularly favorable for the rupture of vessels, a fact on which Little laid stress.

Small capillary hemorrhages within the various tissues of the infant are an almost constant accompaniment of birth. Seitz,<sup>3</sup> in speaking of this fact, refers to an investigation by Paul, who made an ophthalmoscopic examination on 200 newborn children, and found retinal hemorrhages in a large proportion of cases—in 50 per cent. of those in which the mother's pelvis was contracted, in 40 per cent. when the labor was protracted and complicated, in 40 per cent. when the labor was premature but otherwise normal, and in 20 per cent. of normal labors.

Undue pressure on the child's head, due either to a disproportion between its size and that of the mother's pelvis, or to the faulty application of forceps, may cause direct injury to the brain, or, as is much more common, result in intracranial hemorrhage. The bleeding is usually from a rupture of one of the venous sinuses, or of veins emptying into them. The vessels can be injured by the bones of the skull being forced one under the other, or by being implicated in a tear of the tentorium.

Beneke<sup>4</sup> and a number of others have pointed out how frequently the tentorium is torn during difficult labor, due, they think, to stretching of the dura caused by compression of the head. They regard this as by far the most important cause of the birth hemorrhages. Seitz<sup>5</sup> although confirming the frequent occurrence of injury to the tentorium, insists that this can account for at most but half of the cases, and cannot be the cause of clots confined to the convexity of the hemispheres, and which are due, without doubt, to rupture of the longitudinal sinuses or its veins.

Some idea of the frequency of these meningeal hemorrhages during birth can be gathered from the records of autopsies done on stillborn babies and on those which died shortly after birth. Peterson<sup>6</sup> refers to

<sup>1</sup> *Trans. Obstet. Soc. London*, 1861, iii, 293; also *Treatise on Deformities*, 1853.

<sup>2</sup> *Am. Jour. Med. Sc.*, 1885, clxxvii, 58; *Am. Jour. Obst.*, 1885, xviii, 73.

<sup>3</sup> von Winkel, *Handbuch der Geburts.*, 1907, iii, 49.

<sup>4</sup> *München. med. Wchnschr.*, October 11, 1910, p. 2125.

<sup>5</sup> *Zentrabl. f. Gynäk.*, 1912, xxxvi, 1.

<sup>6</sup> *Nervous and Mental Diseases*, Church and Peterson, 1908, p. 894.

the figures given by Litzmann, who found 35 cases of meningeal hemorrhage among 161 stillborn children. Parrot, in 34 autopsies of the newborn, found 5 with blood in the arachnoid cavity and 26 with hemorrhage into the subdural space. H. R. Spencer, in 130 autopsies on stillborn children, found 53 instances of hemorrhage from the pia and arachnoid.

Holt and Babbitt<sup>1</sup> in their analysis of 10,000 confinements at the Sloan Hospital in New York, found among 9318 living births that 291 infants died during the first two weeks. The cause of death was given as intracranial hemorrhage in 19 cases, and as injury to the head in 10. The 429 stillbirths were not analyzed in this relation, but among the 196 ascribed to accidents of labor, many intracranial hemorrhages must have occurred.

Although the hemorrhages occurring during labor are in the great majority of cases meningeal, they do occur at times within the brain substance. Weyhe, in an analysis of the records of the Pathological Institute at Kiel, in 959 autopsies in young infants found 122 cases in which intracranial hemorrhages had occurred. In many cases there were hemorrhages in more than one location. A clot was found 80 times in connection with the dura and 56 times with the arachnoid; in 35 instances it occurred within the substance of the brain, and in 21 within the ventricles. There were 32 instances of relatively large hemorrhages; 5 of these large hemorrhages were intracerebral, and in all but one case it seems fair to assume that the rupture took place at birth.

**Postnatal Cases.**—Hemiplegia in children due to causes effective after birth may occur at any age, but is much more frequent in the first two years. This is well shown by Table IV, which is compiled from that given by Freud,<sup>2</sup> with the addition of the writer's collected cases. Of the 607 cases recorded in the first decade, 344 (56.7 per cent.) were in the first two years, and 516 (85 per cent.) in the first half of the decade.

TABLE IV.

	Congenital.	Up to 1.	Up to 2.	Up to 3.	Up to 4.	Up to 5.	5 to 10.
Osler . . . . .	15	45	22	14	1	3	9
Wallenberg . . . . .	19	35	29	17	9	9	30
Gaudard . . . . .	11	17	7	5	8	2	14
Lovett . . . . .	7	5	12	..	..	..	..
Strümpell . . . . .	..	7	8	4	5	..	..
Sachs and Peterson . . . . .	22	27	17	16	4	4	8
Gowers . . . . .	..	23	23	14	10	..	10
Freud-Rie . . . . .	2	3	11	9	3	3	2
J. H. H. cases . . . . .	32	16	37	17	8	7	18
Total . . . . .	108	178	166	96	48	28	91

The mortality statistics of the *United States Census Report* for 1906 show the same thing more strikingly, for of the 961 deaths recorded as due to apoplexy and paralysis in the first decade, 882, or nearly 92 per

<sup>1</sup> *Jour. Am. Med. Assn.*, 1915, lxix, 287.

<sup>2</sup> *Die Infantile Cerebrallähmung*, Nothnagel, 1897, p. 44.

cent., occurred in the first five years. Although these figures are exclusive of stillborn children, they probably include many children who died shortly after birth from accidents incident to it, for 621 died within the first year and only 127 in the second.

It is also probable that a considerable number of the cases of hemiplegia included in Freud's table as developing in the first and second years were really congenital. For instance, of the 16 Hopkins cases in the first year, the histories of 5 merely stated that the paralysis was noticed at varying times during the infancy: once when the child was one month old, once when four months, twice when six months, and once when eight months; in none of these cases could the parents be sure that the condition had not existed since birth. In 5 of the 37 cases in the second year the disability had become apparent when the time arrived for the children to begin to walk, and here again no definite onset could be determined, so that these 10 cases might fairly have been added to the 32 congenital ones.

**Etiology.**—The cerebral changes which underlie the hemiplegias developing after birth and during infancy are, in general, the same as those which are present at later periods of life, but are influenced by special factors. The vascular system of a child is much less apt to show permanent changes, and on this account the spontaneous rupture or the occlusion of a bloodvessel is less apt to occur. On the other hand, acute infections seem especially prone to affect the brains of the young. Table V represents the etiology of the cases in the first decade. These are distributed by years in the first half of the decade and in the last half are considered together.

TABLE V.  
Congenital, 32; probably congenital, 10; total, 42.

Postnatal cases.	Year of onset.						Total
	1	2	3	4	5	5 to 10	
Acute infectious diseases:							
Whooping cough . . . . .	1	1	1	1	1	1	6
Typhoid fever . . . . .	..	1	1	..	1	2	5
Measles . . . . .	..	2	..	1	..	1	4
Diphtheria . . . . .	..	1	..	..	..	3	4
"Sore throat" . . . . .	..	..	..	..	1	..	1
Croup . . . . .	..	..	..	1	..	..	1
Gastro-intestinal trouble . . . . .	..	1	1	..	..	1	3
Marasmus . . . . .	1	..	..	..	..	..	1
Pneumonia . . . . .	..	1	..	..	..	..	1
Vaccination . . . . .	..	1	..	..	..	..	1
"Fever, brain fever," etc. . . . .	4	4	3	1	1	2	15
After convulsion without other cause:							
With marked brain involvement . . . . .	1	3	1	..	..	..	5
Convulsions at onset . . . . .	2	12	3	1	..	1	19
Developed in course of epilepsy . . . . .	..	..	1	..	..	..	1
Congenital syphilis . . . . .	..	..	1	1	2	..	4
Brain tumor . . . . .	..	..	..	..	..	3	3
Trauma . . . . .	..	3	2	1	..	..	6
Heart disease . . . . .	1	..	..	..	..	1	2
Poisoning . . . . .	1	..	..	..	..	..	1
Hysteria . . . . .	..	..	..	..	..	1	1
Without determined cause . . . . .	..	2	3	1	1	2	9
Total . . . . .	11	32	17	8	7	18	93

*Hemorrhage.*—The rarity of intracerebral hemorrhage in childhood is commented on by most writers. Meningeal hemorrhages are more common, but even these, when not associated with trauma or with one of the acute infectious diseases, are rare. Sachs<sup>1</sup> thinks that hemorrhage occurs more frequently than most authors admit, and seems to regard it as the most common cause of these paralyses, even of the cases that develop after birth. The bleeding, he believes, is from a cortical vessel, although it may be from a vessel within the depths of the brain.

Alterations in the vascular walls may occur in childhood as an early degeneration, or more frequently as the result of some acute infection. Sachs cites von Recklinghausen's statement that fatty degeneration of the cerebral vessel is not uncommon in children. The rupture of such a diseased vessel must account for some exceptional cases of apoplexy of the first decade, and a very few of the writer's cases may best be so explained. However, there is no record in the Johns Hopkins Hospital of the occurrence of a postnatal intracerebral hemorrhage in a child under ten years. This hospital admits relatively few children, and the rarity of such cases can be better judged from other records. Holt says that he has seen but one case. Among 830 autopsies at the Babies' Hospital in New York, the records of which the Staff assisted me in examining, there is no such case, and Dr. John Howland informed me that in 1416 autopsies at the Foundling Hospital there was just one intracerebral hemorrhage. Sidney Phillips<sup>2</sup> reports the occurrence of a spontaneous hemorrhage into the pons Varolii of a girl, aged fourteen months. The child, who was apparently well when put to bed at 3 P.M., was found comatose an hour afterward, and remained so for the fourteen days which she lived. No cause for the hemorrhage was discovered.

Among the 93 postnatal cases there are 9 classed as developing without known cause, *i. e.*, so far as could be discovered; the paralysis developed more or less suddenly in children who were not sick at the time, very much as we are in the habit of seeing an apoplexy occur in the late decades. The history of the onset in five of these cases is so incomplete that some or all of them may well belong to other classes. Two of the four remaining cases are of particular interest. The first was a girl, aged six years, who, while in apparent health, fell, paralyzed on her right side, but was not unconscious for any considerable time. The paralysis improved, but intense athetosis followed in the arm, for which she was admitted on six different occasions, the last time when she was twenty-five. The other case occurred in a girl, aged nine years. She was at her desk in school when she fell unconscious, and remained in a more or less dazed condition for nearly two weeks. The paralysis was on the left side and cleared up well. When twelve she had a second attack, which followed a right-sided headache, and came on while she was walking around her room. She fell, paralyzed again on her left side, but without loss of consciousness. The power again returned, but rather more slowly and athetosis followed. The onset in these cases was so characteristic of a vascular lesion that it was difficult to think of any other explanation.

<sup>1</sup> *The Nervous Diseases of Children*, second edition, 1905.

<sup>2</sup> *Lancet*, 1909, i, 1680.

No sources of emboli were found, and we have been led in each case to assume the occurrence of either a rupture or a thrombus of one of the arteries in the substance of the brain.

In the other two cases the paralysis came on at the eighteenth or nineteenth month. In one case the onset was sudden; the baby was unconscious for ten days and was paralyzed on the right side. In the other case the onset was rather gradual, in the course of one or two days.

*Hemorrhage* has been the assigned pathological condition in many of the cases of hemiplegia associated with definite etiological factors. This would seem justified in certain of the cases developing after injury to the head when the skull had not been fractured, although the possibility of an encephalitis must not be forgotten. With fracture of the skull, hemorrhages, particularly from the meninges, are common, no matter at what time of life the accident occurs, and even when the fracture is not apparent it cannot always be excluded. In only one of the traumatic cases was the skull known to be injured, and in that there was a perforating wound.<sup>1</sup> In one case the fall may have been merely coincident with the attack of paralysis. This was in a girl, aged two years, who had been sick with some fever and who fell from a little cart. When picked up she was paralyzed on the left side. In the other four cases the falls were of sufficient gravity not to be overlooked as etiological factors.

Cerebral hemorrhage during infantile convulsions has been assumed as the cause of many of the early hemiplegias. The fact of the sequence of convulsions and paralysis is most striking, and the supposition that blood-pressure changes, incident to the fit, may have caused rupture of the bloodvessels is attractive. That the vessel walls, on the other hand, are in the vast majority of cases amply able to withstand any increase of pressure incident to the most severe convulsions, is shown by the rarity of their rupture in cases of epilepsy. Capillary hemorrhages are, indeed, frequently found in patients dying in the status epilepticus, but these do not appear to be large enough to account for paralytic symptoms. In none of the cases of this series did the paralysis follow a fit in an epileptic, unless two cases in the first decade may be so considered. The more striking of these was that of a hemiplegic idiot seen at seven, who began to suffer from convulsions at eight months, which recurred every two or three weeks. When two years old the patient had a severe attack lasting four hours, after which his mother noticed that he was paralyzed on the left side. He did not walk again for more than a year. A single convulsion recurred six months after the attack of hemiplegia, and there was then an interval of four and a half years, after which he had numerous seizures.

Including these cases, there were 54 of the 93 postnatal cases in which the paralysis was ushered in by convulsions, but in no other of these was there any record of the child having had convulsions previously. Among these 54 cases many developed in association with various etiological factors; 14 with acute infectious diseases and 10 from a variety of causes, congenital syphilis, brain tumor, trauma, etc., and the primary cause of the cerebral lesion must have varied much—hemorrhage, thrombosis,

<sup>1</sup> Had I analyzed the surgical records, without doubt many more cases of infantile hemiplegia due to trauma would have been added.

embolism, and inflammation. In the remaining 30 the cerebral involvement, as judged by the sudden onset of the convulsions in apparent health, seems to have been the primary lesion. In 14 of these there was either more or less prolonged fever, or definite evidence by protracted coma, etc., to show that the brain was seriously involved from the first. The discussion as to the pathology of infantile hemiplegia has, in a great measure, centred around cases similar to these, *i. e.*, those cases in which the paralysis developed acutely in apparently healthy children, with more or less fever and evidence of involvement of the brain. The convulsions are either complete or, for the most part, unilateral, and one must assume a cause that acts more or less locally from the first, and it is obviously unsatisfactory to explain the paralysis as the result of the convulsions without giving an adequate cause for these, especially as there is reason to doubt the efficiency of convulsions in themselves to cause permanent gross lesions. Gowers assumes, as the most satisfactory explanation of these cases, "a sudden occlusion of a small surface vein by clot, with the consequent intense congestion and hemorrhagic softening of the region of the cortex."

The many points of similarity between the onset of these cases of infantile cerebral hemiplegia and those of acute poliomyelitis led Strümpell, in 1884, to assume as their anatomical basis a lesion in the brain analogous to that in the cord, which was believed to underlie infantile spinal paralysis, *i. e.*, an inflammation confined to the gray matter of the motor cortex of the brain (polioencephalitis) corresponding to the inflammation limited to the ventral horns in the cord. This theory was advanced at a time when the anatomical data in regard to spinal infantile paralysis were very limited and when there were practically none concerning inflammation of the brain, so that in spite of its great attraction it did not receive general acceptance. Since then a great deal of work has been done on the inflammatory conditions of the central nervous system, and the views in regard to these conditions have been very much broadened. We now believe that infantile spinal paralysis is due to a specific organism which manifests itself for the most part by a diffuse inflammation about the vessels of the meninges and central nervous system, involving particularly, but by no means exclusively, the anterior gray matter of the spinal cord.

Many cases of infantile hemiplegia, which develop acutely with fever, convulsions, etc., and which are presumably due to an encephalitis, are independent of any such diseases. It is an attractive and obvious thought that certain of these cases may be due to a specific organism, analogous to, if not identical with, that of poliomyelitis. That the brain is involved in many cases of poliomyelitis is manifest clinically, and in practically every fatal case some inflammation of the pia mater at the base of the brain can be demonstrated.

However, an encephalitis of the degree that would entail a hemiplegia seems extremely uncommon, for very few such cases have been reported during the recent epidemic of poliomyelitis. At present then we should regard the organism of poliomyelitis as only one of the causes, and probably a rare one, of infantile hemiplegia.



Among the 93 postnatal cases there were 26 instances in which the paralysis developed in association with one of the acute infectious diseases, 6 in relation to whooping cough, 5 with typhoid fever, 4 with measles, 4 with diphtheria, 3 with gastro-intestinal disturbances, and the other 4 distributed among various conditions. The cerebral lesions in these cases are by no means always the same. Inflammatory processes must account for a number of them, but hemorrhage, thrombosis, and embolism have been found with varying frequency.

*Whooping cough* is accountable for more intracranial hemorrhages than any of the other conditions. A great increase in venous pressure during a paroxysm of coughing is the usual explanation, but Neurath believes that changes in the vessel wall are more important than increased pressure. The hemorrhage is thought to be from a meningeal vein. Rhein, who analyzed the autopsy findings of 21 cases of pertussis, found the hemorrhage when present most frequently in the substance of the brain. In his case the lesion was inflammatory, and Neurath refers to a number of such cases and believes that hemorrhage is less common than is supposed.

*Typhoid fever* we believe to be accountable for 5 of our cases, although this seems a large number when we consider the relatively few cases of hemiplegia associated with this disease. Smithies was able to collect only 40 cases. The age of 32 of these cases is interesting; 9 were in the first decade, 8 in the second, 13 in the third, and only 2 beyond that time. When one considers how relatively infrequent typhoid fever is in the first decade, it is surely remarkable to find more than one-quarter of the cases of hemiplegia occurring in children under ten. Smithies was able to find the record of 5 autopsies, only one of which was in a case in a child, and the examination was made twelve years after the onset of the hemiplegia. Two of the other 4 cases occurred among our records, and have been reported by Osler. In all of the cases occlusion of an artery was found, so that from the evidence at hand we must conclude that cerebral thrombosis is the cause of most cases of hemiplegia developing during typhoid fever. It is probably secondary to changes in the artery walls.

*Diphtheria* is accountable for 4 of our cases, or, indeed, 5, if we include the case in which the history simply recorded a severe sore throat. If hemorrhage is the most common cause of the hemiplegias associated with pertussis, and thrombosis the most common cause in relation to typhoid fever, it seems that embolism plays this part in the hemiplegias developing during diphtheria. Thus, Rolleston,<sup>1</sup> in his collection of 65 cases, found 15 which were completed by autopsies. Among these, hemorrhage was found in 1, thrombosis in 2, embolism in 10, embolism and thrombosis in 1, and atrophy of one hemisphere in 1 case.

*Measles* is represented in this list by 4 cases, but there is not any case which occurred in association with *scarlet fever*. Concerning this latter disease, Rolleston<sup>2</sup> speaks of the rarity of hemiplegia in relation to it, but he was able to find the records of 57 cases, 8 with autopsy. He concludes that it is usually due to cerebral embolism, but may follow

<sup>1</sup> *Rev. Neurol. and Psychiat.*, 1905, p. 722.

<sup>2</sup> *Ibid.*, 1908, p. 530.

thrombosis, hemorrhage, or acute encephalitis. Under this last heading he thinks uremic hemiplegia may probably be placed.

Except in the instance of encephalitis, the involvement of the brain during the course of an acute infectious disease is secondary to involvement of the vascular system. The association between emboli and endocarditis is evident, and that the vessel walls may be involved in children during the course of acute infectious diseases has been shown by a number of authors, particularly by Wiesel,<sup>1</sup> and that the occurrence of thrombosis is secondary to this involvement cannot be doubted. The tendency to rupture of an artery, as in whooping cough, is probably brought about by the weakening of the vessel wall through this process. This arterial disease appears in most instances to be entirely recovered from, but it does at times leave the artery wall permanently damaged, and so acute infectious disease during infancy may be the predisposing cause of later vascular accidents.

*Embolism* is not uncommon in association with acute endocarditis occurring in the course of the acute infectious diseases (diphtheria, scarlet fever, etc.), but it is remarkable how rarely it occurs in chorea, a disease in which the heart is so generally affected. There is but one case in the series, although we have seen a large number of cases of chorea with involvement of the heart. The case was that of a little girl, aged ten years, who had had chorea a year and a half before, which was followed in the next year by rheumatic fever and endocarditis. She had been up about four weeks after this attack, when she fell from a bench, had repeated convulsions, and was paralyzed on the right side with loss of speech. The only other case in which it seems reasonably certain that the hemiplegia was due to an embolus was in a little child, aged one year, who had congenital abnormality of the heart. This organ was on the right side. The sounds at the apex were clear, but over the right second costal cartilage there was a grating systolic murmur. Four weeks before we saw the patient the mother discovered, on taking him up in the morning, that his right arm and right leg were completely paralyzed, the attack having come on during sleep.

The importance of *hereditary syphilis* in the causation of infantile hemiplegia has given rise to a great deal of discussion. Sachs believes that it has very little influence, but other authors give it prominence. In 4 cases of this series it appeared to be the most likely etiological factor and in 1 case there was hardly a doubt. This was in a child whose brother and sister had been treated for congenital syphilis, and who, himself, when twenty-one months old, had marked manifestations of the disease. When four years old he had an attack of right hemiplegia, with loss of speech, choked disk, and retinal hemorrhages. He was at this time passing through whooping cough, but this case is not included among those occurring in whooping cough. He improved under specific treatment, but the left optic nerve became atrophied. When five years old he had a second attack, this time on the left side, and speech was again affected. The paralysis improved, but there was a marked mental

<sup>1</sup> *Ztschr. f. Heilk.*, 1906, xxvii, 262.

defect. There was another case in which the history seems clear. The father had syphilis, and a specific eruption developed in the baby when it was two months old. The child was given antisyphilitic treatment. When four years old convulsions developed, followed by right-sided paralysis. When seven the child had a painful swelling of the anterior border of the tibia, which disappeared under specific treatment. The vascular lesion in these cases was presumed to be an occlusion, secondary to syphilitic changes in the vessel walls.

During the course of *brain tumor* hemiplegia is not uncommon, and three such cases occurred on the medical side; but in these the paralysis was of rather slow development, and so probably did not depend upon a hemorrhage into the substance of the tumor.

*Hysteria* may cause a hemiplegia at almost any age, and we have the record of one case, that of a little girl seen in her tenth year, suffering from her second attack. The first attack had been a year before. She had paralysis of the right arm and leg with typical sensory disturbances. The disability cleared up at once under suggestion and faradic stimulation.

**Cerebral Diplegia.**—Closely allied to the hemiplegias which develop early in the first decade are the infantile cerebral diplegias. The same etiological factors, congenital abnormality of the brain, injury to the head during birth, hereditary syphilis, inflammatory conditions, etc., apply to both. In the one case the lesion is unilateral, while in the other both sides of the brain are affected. The two sides of the brain may be unequally involved, and it is not infrequent to find one side of the body more affected than the other, and only portions of the hemisphere may be injured and corresponding symptoms result. Not infrequently the upper portions of both hemispheres are most involved, and the symptoms are largely confined to the legs (cerebral spastic paraplegia). The lesions occurring before or at the time of birth are more frequently bilateral, and diplegia is the common result of these processes. Later in the decade the reverse is true, and hemiplegia is then much more frequent.

Among 100 cases of infantile cerebral palsies in the Neurological Department of the Johns Hopkins Dispensary, and in which the process was definitely congenital, 68 were diplegia and 32 hemiplegia. There are records of only 10 cases of diplegia which developed after birth. Five of these were in the first year, 3 in association with acute illnesses, measles, pneumonia, whooping cough, and the others with indefinite illnesses. Of those developing after the first year, 2 were with fever, 2 are said to have followed falls, and in 1 no cause could be given. Besides these 78 cases, there were 4 others in which the early history was unknown, and in all the processes may have been congenital.

The various *symptoms* of infantile spastic diplegia (Little's disease) are, for the most part, those of hemiplegia, which will be described in detail later, only in this case both sides of the body are involved. Certain symptoms are so pronounced that they require special mention at this time. These are particularly those depending upon the lack of development of the brain, for it is obvious that when the brain has been severely injured early in life it cannot develop normally, and so these children show every degree of idiocy. The degree of mental retardation is, in a

general way, proportionate to the motor defect, but not always so, and the intelligence of certain patients who have never been able to walk or even to sit up is surprising. It has been suggested that in such cases the lesion is not in the brain itself but in the upper part of the spinal cord. The disparity is more common in the other direction, and one often sees children who show only a slight degree of spastic paralysis, but whose mentality is very low. Indeed, every combination occurs.

As in certain cases of congenital hemiplegia, the disability may not be noticed until it is time for the child to begin to walk, but usually there are symptoms which should attract attention. Directly after the difficult birth there is often trouble in establishing respiration, and not infrequently the child has muscular twitchings or even convulsions. The increased intracranial pressure may be shown by a tense non-pulsating fontanel and a general lack of animation, a higher grade of choked disk than usual, and death may occur rapidly. In less severe cases the baby seems to recover and to perform all of its functions normally. These, it must be remembered, are largely reflex, depending on the lower centres. Later, when the time arrives for the baby to show certain spontaneous activities, as holding up its head, sitting up, learning to walk, and to talk, the parents are distressed to see that these are not acquired. The spastic condition of the legs is often first noticed when the child is several months old by a resistance in the adductor muscles as the mother changes the child's napkins. When the child is held in the standing position this adductor spasm comes out more clearly and the legs are held in extension and firmly together. Indeed, at times the spasticity can be brought out only in this manner. In other cases it is so pronounced that it attracts attention very early (congenital spastic rigidity). Such babies become rigid upon the slightest cause, when handled or when startled by a noise.

Even when the spasticity is not excessive these children learn to walk late; not usually before the third year, and often even much later. The walk is awkward and spastic, and frequently shows the typical crossed leg progression, each leg as it is advanced being thrown across the other by the strong contraction of the adductor muscles. The finer movements of the arms and hands are learned, if at all, with difficulty, and even the coarser movements are performed with marked incoördination. Such children may be entirely unable to learn to talk, and when they do, the articulation is apt to be very inexact, even when the mental development is of such a grade as to make speech possible.

In infantile cerebral palsies, both of the hemiplegic and diplegic type, abnormal spontaneous movements are common; indeed, they are rarely seen in cases which develop later in life. The characteristics of these conditions, athetosis, postparalytic chorea, etc., will be discussed later. Athetosis in its classical form occurs in diplegias, and the condition described as double athetosis is confined to this state. Lewandowsky<sup>1</sup> insists that double athetosis is not simply the involvement of both sides of the body with typical athetosis, but that it is a different condition and depends upon excessive associated movements. Such patients may

<sup>1</sup> *Deutsche Ztschr. f. Nervenhe.*, 1905, xxix, 339.

be quite quiet when undisturbed, but when their attention is attracted, or when they endeavor to perform any voluntary act, the muscles of the face are thrown into the most startling grimaces, the arms are moved about in quick excessive choreiform movements, which suggest strongly those seen in Huntington's chorea. Double athetosis may occur in its most pronounced form when there is but little motor paralysis.

*Epilepsy* not infrequently develops, although perhaps this is somewhat more common in the hemiplegic than in the diplegic type. The affected limbs may be retarded in their growth, and the whole physical development be stunted. This is not always so, and even in a severe case the body may reach its normal growth.

**Decade II.**—Fewer cases of hemiplegia occur in the second decade than at any other time of life, except extreme old age, and even here the liability to these vascular accidents is much greater. The difference between the first and second decades depends largely upon the great number of cases in the first three years of life due to accidents at birth, and the liability of the infant's brain to inflammatory processes. The 25 cases falling in this decade were divided as follows: 6 were due to trauma, and 6 occurred in the course of acute illness, diphtheria 2, meningitis 2, typhoid fever 1, and undetermined illness 1. One other case, that of athetosis, developed after a disease that was said to be meningitis. Two were in relation to brain tumors and 2 were hysterical. The remaining 8 cases are of particular interest, for they developed suddenly in apparent health, and presumably were due to primary vascular lesions. Similar cases occurred in the first decade, but their relative frequency is much greater in the second. This is what would be expected from the increasing number of individuals who must have damaged vascular systems from congenital weakness, acute illnesses, etc. As none of the cases was completed by autopsy, the pathological basis for the paralysis must be largely a matter of inference.

*Embolism.*—Embolism seems almost surely to account for 3 of the 8 cases. The first was in a girl, aged eleven years, who had frequent attacks of sore throat and arthritis. Her paralysis came on suddenly while she was at work in the kitchen; her left leg gave way so suddenly that she fell on the stove. She did not lose consciousness, and was able to keep about for a few hours, but the next morning she noticed that her left arm and leg were weak. There was a typical left-sided hemiplegia, and the examination of the heart revealed stenosis and insufficiency of the mitral valve. The second case was quite similar. The third case was that of a colored boy, aged thirteen years, who was awakened early one morning be intense pain in the stomach, vomited, had to be assisted back to bed on account of staggering, went to sleep, but was awakened three hours later by another attack of vomiting, after which he became unconscious and was brought to the hospital the same day. He was comatose, the right arm and leg were motionless, and the left arm and leg were in almost constant, restless movement. The head was turned to the right, and the right leg was stiff. There was mitral insufficiency. The ophthalmoscopic examination showed evidences of increased intracranial pressure, and a decompression was done. Dr. Cushing found the left side of the

brain red and dry, resembling a hemorrhagic infarct. The boy recovered with right hemiplegia and aphasia.<sup>1</sup>

*Hemorrhage.*—This appears to account for at least one of the 5 remaining cases, which was in a girl, aged sixteen years, who, since she was nine, had been seen a number of times suffering from attacks of Henoch's purpura. While on her way home from a friend's house she had difficulty in speaking, and that evening found it hard to eat. There was, however, no weakness of the limbs until the next morning, when the right arm was affected. On the second day she had convulsions, and was admitted to the hospital aphasic and weak on the right side. She had recurrent convulsions, beginning in the right hand; the head and eyes turned to the right, the right leg was next involved, and the convulsions then became general. After each convulsion she was more comatose, and new purpuric areas appeared on various parts of the body. Dr. Cushing operated and found the dura tense and the brain and subpial space of a uniform cherry-red color. Nothing but decompression was attempted. The patient died on the day after the operation, and four days after the onset of the cerebral symptoms. No autopsy was permitted.

Hemorrhage may have accounted for the other 4 cases, although thrombosis cannot be excluded. In none of them could a source for an embolus be found. They occurred suddenly, in 2 instances the paralysis being discovered in the morning, after an undisturbed sleep, and one case, a girl, aged fourteen years, while working in a very hot schoolroom, felt nervous and fell unconscious on her desk. This was followed by paralysis on her right side with speech disturbance. The examination nine months afterward showed the signs of a typical organic hemiplegia. In this case the heart was normal and no arteriosclerosis was discovered.

A most interesting specimen was brought to the Pathological Department. This was the brain of a school boy, aged fifteen years, who seemed perfectly well upon going to bed. At 1 A.M. he aroused his room-mate by loud screams, and complained of intense pain in the head. He quickly became unconscious, and died at 8 A.M. The autopsy showed an enlarged thymus, lymphatic hyperplasia of the intestine, lymph glands, and spleen, with oedema and congestion of the lungs. The brain showed a large hemorrhage in the right temporal lobe. A careful microscopic examination failed to reveal any disease of the cerebral vessels.

The cases which were associated with acute illnesses differ only from those occurring in the first decade in the age of onset, except the two cases associated with meningitis. In one of these the type of meningitis could not be determined, but the other case was proved by lumbar puncture to be acute cerebrospinal meningitis. Hemiplegia is a rare complication of epidemic cerebrospinal meningitis. Castaigne and Rivet<sup>2</sup> report a case in a boy, aged seventeen years, and could find the record of but one other case. They ascribe the complication to encephalitis.

<sup>1</sup> In connection with these three cases I may refer to a case already reported in *The Johns Hopkins Hosp. Bull.*, 1901, vol. xii, of embolism of the central artery of the retina in a girl, aged sixteen years, who had chorea and mitral insufficiency.

<sup>2</sup> *Bull. et mêm. Soc. méd. d. hôp. de Paris*, 1909, xxvii, 900.

**Decade III.**—The liability to apoplexy in the third decade is about three times as great as in the second. This is due to the influence of syphilis and to the increased number of cases depending upon cardiovascular changes. Pregnancy and the puerperal state account for a certain number of cases, and a few are in association with various pathological conditions (pernicious anemia, hemophilia, etc.).

*Syphilis* was recorded as having occurred in 20 of 62 cases of this series, and there can be no doubt that to this number others should be added. Of 12 doubtful cases, 6 were in negroes—5 men and 1 woman—in whom syphilis is extremely prevalent, but in whom it is always difficult to get a history of the infection, and there were 2 white women, both of whom were probably syphilitic, for one had many miscarriages and the other was a prostitute. The cerebral complication may follow the primary lesion quickly, indeed within the first year, or it may be delayed for a long time, ten or more years. One case occurred at five and one at seven months, and 4 others at about one year after the infection. The average time was approximately four and a half years.

*Thrombosis.*—Thrombosis with consequent softening of the nervous tissue, is the lesion upon which practically all the cases of apoplexy due to syphilis depend. This depends upon a specific change in the artery walls, the so-called syphilitic arteritis. Under thrombosis should probably be grouped most of the cases occurring with acute infectious diseases, and the cases occurring shortly after childbirth. The hospital list contains 5 of the former and 3 of the latter in this decade. Of the 5 cases occurring during the course of an acute infection, 4 were with typhoid fever, and one with pneumonia. Two of the typhoid fever cases occurred in the hospital, and both died. The autopsy on one revealed thrombosis of branches of the middle cerebral artery. The case which developed during the second week of a severe double pneumonia was possibly due to an embolus, for when the patient was examined six months later his heart showed evidences of serious organic disease. Hemiplegia is an extremely rare complication in pneumonia.

Hemiplegia and other focal manifestations of cerebral lesions occur in association with *pregnancy* and *childbirth*. These cases naturally fall in the third and fourth decades, and in this series 3 of the hemiplegias of the third decade were of this character. They developed after childbirth, one on the fourth day, one on the ninth, and one a month afterward.

The lesions in such cases vary a great deal and hemorrhage, thrombosis, and embolism have all been found. von Hosslin<sup>1</sup> has reviewed this subject. Hemorrhage is the most common lesion, particularly in those cases associated with marked albuminuria or symptoms of eclampsia. In this association, a number of cases have been reported in which the brain showed only œdema.

Women with diseased arteries from any cause run special risks during pregnancy and confinement. An illustration of this was a woman who died in the obstetrical ward. She was thirty-five years of age, and when thirty-two had a left-sided hemiplegia, which had no association

<sup>1</sup> *Arch. f. Psychiat.*, 1904, xxxviii, 730, and 1905, xl, 445.

with pregnancy. When thirty-five she became pregnant, and at the eighth month she fell unconscious, apparently in uremia. She was brought to the hospital, delivered immediately, but died soon after. Autopsy showed a softened area due to the old lesion, and a large hemorrhage into the pons Varolii. The arteries at the base showed marked sclerosis. The liver did not show the changes characteristic of eclampsia. Hemorrhage does occur during parturition in women whose arteries show no general disease.

*Embolism.*—von Hosslin found embolism the least common cause of apoplexy during pregnancy, although Meyer-Ruegg believes that it is the most common. The source of the emboli is endocarditis. This may be acute, developing spontaneously, or as an exacerbation of a chronic process; or a chronic endocarditis may be the source of an embolus. Embolic occlusion of a cerebral vessel is the reasonable explanation of the 12 cases of hemiplegia which occurred between the twentieth and the thirtieth year in individuals suffering from organic heart disease. If syphilis be excluded this is by far the most common cause of the hemiplegias in the third and fourth decades. Three of these 12 cases developed while in the hospital; 2 in association with ulcerative endocarditis, and the third in a case of rheumatic fever with cardiac involvement. The other 9 cases occurred while the patients were about their usual vocations, and when the cardiac condition had become chronic.

*Hemorrhage.*—This is uncommon before the fifth decade, but even though syphilis may account for the majority of the 12 cases in the third decade, in which no cause could be determined, still some of them are hard to explain in this manner. For instance, a man, aged twenty-one years, who admitted only one exposure to venereal infection and who denied all knowledge of any such accident, fell while working and was paralyzed on his right side. A physical examination revealed no lesion of the heart, and in such cases, without an autopsy, it is impossible to exclude cerebral hemorrhage. However, in 3 of the 62 cases occurring in this decade, there can be little doubt that cerebral hemorrhage did occur, and in one this was proved by autopsy. The patient was a woman, aged twenty-two years, who had nephritis, and who died eleven days after an operation for decompression. At autopsy a large capsular hemorrhage was found. The other two cases were associated with conditions which make hemorrhage almost certain. In one of these, a white man, aged twenty-eight years, with pernicious anemia, a right hemiplegia appeared nineteen days before death. The autopsy revealed hemorrhages in the various organs of the body, but unfortunately the brain was not examined. The other case was in a hospital orderly who came from a family of bleeders, and who was himself a hemophiliac.

*Trauma.*—Trauma accounts for 2 of our 62 cases. One of these is of interest, as the head was not directly affected. The patient, a brakeman, aged twenty-one years, was injured while coupling cars; he was crushed about the chest and right hemiplegia developed, which may have been due to a hemorrhage secondary to great venous congestion.

*Hysteria.*—This accounted for 5 of hemiplegias in the third decade. Four of these were in women and one was in a man.



**Decade IV.**—In this decade the liability to vascular lesions is again greatly increased, being about three times as great as that in the third decade. The influence of syphilis is still marked in the fourth. Of the 102 cases, 32 gave a definite history of syphilis, which almost equals that in the third decade, being 31.3 per cent. in the former and 32.2 per cent. in the latter. But the group of cases in which no definite cause could be determined is much larger, making 44.1 per cent. In this group many cases must be included which are due to syphilis, although this has not been recognized or not recorded. As other conditions which cause arterial degeneration are constantly increasing with advancing age, the proportion due to syphilis naturally becomes smaller as the age becomes greater, and, therefore, in this decade, this group must contain a relatively smaller number of unrecognized syphilitic cases.

The time between the syphilitic infection and the occurrence of the apoplexy in the cases which fall in the fourth decade is, as would be expected, somewhat longer than that which was found for the cases of the third decade. In this series, as accurately as could be determined, it was a little more than eight years. Of the 32 cases, all but 1 occurred in men. This, of course, does not mean that the condition does not occur in women, but only illustrates how difficult it is to obtain a syphilitic history in women.

Acute infectious disease accounts for only 2 cases among the 102 in this decade. In these the hemiplegia developed during typhoid fever, and was presumably due to thrombosis of a cerebral artery. In this connection, a private case, not included in this analysis, seen in consultation with Dr. Chrystie, of Bryn Mawr, Pa., may be noted. It occurred in a woman, aged forty-two years, during the course of a protracted fever, the nature of which was never definitely determined, the diagnosis resting between a mild attack of typhoid fever and an unusual influenzal attack. On the fourteenth day the patient had a convulsion affecting the right side, which lasted about eight minutes, and which left her paralyzed on the right side and aphasic. The left eye was blind, due to obliteration of the central retinal artery. In this case the lesion was almost surely a thrombosis of the internal carotid, implicating its ophthalmic branch as well as the middle cerebral artery. The hemiplegic attack had practically no influence on the temperature, which continued to vary between 98.5° and 101° until the twenty-second day of the disease, after which it remained practically normal.

There is one case in the fourth decade which occurred twelve days after the birth of the eighth child in a woman, aged thirty-nine years; it was associated with a normal puerperium. Two other interesting cases were seen in private practice.<sup>1</sup> These were associated with evidences of toxemia, and both had hemianopia. In one there was a transient left hemiplegia with permanent sensory disturbances. Cerebral thrombosis was the most probable lesion in these cases.

Bilateral capsular softening, due to thrombosis, was found at autopsy

<sup>1</sup> Both of these were seen with Dr. Hiram Woods, and have been reported by him, *Jour. Am. Med. Assn.*, 1908, li, 204.

in a colored woman, aged thirty-nine years, who presented a typical picture of pseudobulbar paralysis.

Embolism, associated with heart disease, was the probable lesion in 11 cases—6 men and 5 women. This was demonstrated by autopsies in 3 cases.

Hemorrhage within the cranium was found in 4 cases at autopsy. Two were in negro men who had had syphilis nine or ten years before death, and are included among the syphilitic cases, although this factor was probably but one of the causes of the vascular disease. They were both markedly alcoholic. In one the intracranial hemorrhage was associated with pachymeningitis and arteriosclerosis, and there was evidence of old cortical hemorrhages. In the other cases there were marked cardiovascular changes and a large clot in the occipital lobe, which had broken through into the subdural space. The other 2 patients were brought to the hospital unconscious, and very little history could be obtained. In one the hemorrhage had taken place in a glioma, which apparently had given no symptoms. The other was in a man, aged thirty-nine years, who had an enormous hemorrhage and general arteriosclerosis.

Spontaneous cerebral hemorrhage is, in fact, generally recognized as being rare before the fifth decade. Von Monakow says that before the fortieth year it is extraordinarily uncommon. However, this lesion was probably present in some of the 45 cases in which the history gave no very definite indication as to the cause of the cerebral lesion. In one case of hemiplegia, which had developed rather gradually, a brain tumor was found at autopsy, and in 2 cases the paralysis was definitely hysterical. Both of these occurred in men.

**Decade V.**—Apoplexy becomes much more common in the fifth decade, the liability to it being three times as great as that in the preceding ten years, and more than twice as many cases occur. In the present analysis there are 137 cases within this period. By the time an individual has reached the age of forty many things may have occurred in his life that make for vascular accidents, and it is often difficult or impossible to determine which has been to blame in a particular case.

*Syphilis*, such an important factor in the third and fourth decades, plays a much less outspoken part in the fifth. There are, indeed, 27 cases in which there is a definite history of a previous infection, but this, in most instances, took place many years before the apoplectic attack, the average time being between sixteen and seventeen years. In many of these 27 cases other conditions were present which may have been effective, and may or may not have been related to the specific infection. This is illustrated by the only case which came to autopsy. The patient, aged forty-seven years, was infected fifteen years previously and had been in the wards on two occasions suffering from symptoms of arteriosclerosis and chronic nephritis. After staying in the hospital three months he became unconscious and a left hemiplegia appeared. The autopsy revealed chronic nephritis, hypertrophy of the heart, acute pericarditis, and thrombosis of the right internal carotid artery.

Cerebral softening was found at autopsy in 7 cases dying between forty and forty-nine. It was not always determined whether the occlusion of

the bloodvessels was due to an embolus or a thrombus, but in one case, that of a man, aged forty-six years, who died during typhoid fever with evidence of left-sided paralysis, there seems to be no doubt that thrombosis of the middle cerebral artery was the cause of the necrotic area in the right internal capsule. This is the oldest case of hemiplegia associated with acute infectious disease of which we have record, and is the only one in the fifth decade. In 2 other cases which were associated with chronic nephritis and arteriosclerosis, the multiple areas of cerebral softening were believed to be thrombotic, and there were numerous cases in which the most probable clinical diagnosis was thrombosis.

In a case of diabetes mellitus a left hemiplegia developed two days before death. At autopsy pulmonary tuberculosis, chronic nephritis, acute endocarditis of the aortic and mitral valves, and infarcts in the spleen and kidney were found. There was slight sclerosis of the vessels at the base of the brain, but no macroscopic lesion was found other than a hyperemic condition of the pia. The probable explanation of the cerebral symptoms was an undetected embolus, which for some reason, an unusual freedom of collateral circulation for instance, had not produced a definite softening. We must also think of the possibility of the case being one of toxic hemiplegia in diabetes mellitus.

*Embolism.*—The heart was noted as definitely affected in 15 cases, and was probably involved much oftener than this. That the hemiplegia was in all these cases due to embolism is, of course, not a just inference, as there were often associated conditions that might have led to thrombosis or hemorrhage. Cerebral hemorrhage was found at autopsy in 4 cases dying in this decade. Two cases of hemiplegia, aged forty and forty-two years, occurred after childbirth.

In 80 cases the hemiplegia developed without determined cause, although in some of them arteriosclerosis was noticed, and evidences of cardiac and renal involvement.

**Decade VI.**—In this period the liability to apoplexy and also the frequency of its occurrence again increases over the fifth decade. It is interesting that the increase in both these relations is almost exactly the same as that of the fifth over the fourth decade, being three times for the first and twice for the second. In the present series, however, there are only 143 cases as against 137 in the preceding ten years. There were 30 deaths against 19 in the fifth decade. In this decade it is more difficult to determine the etiological factors, and 94 cases are included in the group of undetermined etiology. Most of these showed definite evidences of arteriosclerosis, which may have followed a variety of causes. No apology seems necessary for arteriosclerosis at this time of life.

There were 11 cases in which it was definitely stated that the patient had suffered from *syphilis*. This, of course, is an evident understatement of the fact. Men between fifty and sixty have forgotten that they were infected in their early youth, or, believing that such an infection could have no possible bearing on their condition, conceal it; and, on the other hand, the examining physicians, knowing that an early specific infection is apt to be only of slight importance, have not inquired into this feature of the cases as diligently as they should. In these 11 cases

the average time of infection preceded the apoplexy by 21.7 years, and it is probable that in most of them syphilis acted only as one of the causes in producing arteriosclerosis. That syphilis may play its accustomed part at any period of life is shown in 2 of the 11 cases; in one, a man, aged fifty years, a slight hemiplegia appeared while the patient was being treated for tertiary syphilis, he having been infected two and a half years previously. In the other the hemiplegia developed at fifty-five, a year after an undoubted syphilitic infection.

In the only case which came to autopsy there was a history of syphilis twenty years previously. He had marked arteriosclerosis, chronic nephritis, and dilated heart, for which he had been treated in the hospital two months before. He was admitted in a comatose condition, and died in twenty-four hours. An extensive hemorrhage was found in the left cerebral hemisphere, the vessels in the brain being markedly sclerotic. There was no evidence of syphilis at the autopsy.

There were 17 cases in which the heart was noted as being evidently diseased, and in which presumably embolism accounted for the apoplexy. In 4 other cases among 7, in which intracranial softening was found at autopsy, there was a condition of the heart which could have been the source of an embolus. That it is not safe to assume that an apoplexy in an individual who presents the evidence of a cardiac involvement is always due to embolism, is shown by 3 cases dying of intracerebral hemorrhage, in all of which the autopsy showed valvular endocarditis.

Cerebral softening was found in 9 autopsies on patients dying in this decade. Two of these, however, were in cases which had developed in the fifth decade, one due to embolus and one to a thrombus.

*Hemorrhage* was found in 12 cases. Three of these have been referred to as being associated with endocarditis, and one occurred in a syphilitic patient, although there was nothing found at autopsy to suggest this infection. In one other case, in which the history gave no indication of any infection, the autopsy showed what was believed to be a definite syphilitic lesion of the abdominal viscera. In the other cases there was the ordinary vascular degeneration.

**Decade VII.**—In the seventh decade we have 105 cases for analysis, a considerable falling off from the preceding period. The liability, judged from the mortality statistics, is greater, being increased more than two and a half times, and the actual number of registered deaths in Baltimore was 95 for the seventh decade and 69 for the sixth.

At this period it is impossible to make any satisfactory division of the causes which led up to the vascular accident. Fourteen cases did, indeed, give a history of syphilis, but the average time of infection was about thirty years previous, and its influence could have been only problematical. In 2 cases syphilis did seem to have been the predisposing cause. In these, aged sixty and sixty-five years, there was an undoubted infection about three years before the onset of the hemiplegia. In neither case was there any other probable cause determined.

In the great majority of the other cases, evidences of general arteriosclerosis were found, with associated cardiac and renal conditions, states in which thrombosis and hemorrhage are common. In 6 cases the heart

gave such evidences of severe valvular involvement that emboli might have been expected; but in the only case which came to autopsy marked endocarditis, general arteriosclerosis, and a hemorrhage into the left internal capsule were found. Hemorrhage was found at autopsy in 4 cases. In the 2 cases in which cerebral softening was found it was believed to be due to embolism, once from an aneurism of the aorta and once from some unknown source, the embolism being demonstrated on section.

**Decades VIII and IX.**—In the eighth and ninth decades, although the liability to apoplexy continues to increase, the frequency of its occurrence decreases markedly in the last part of this period, owing to the relatively small number of individuals alive between eighty and ninety. In the present analysis there are 25 cases in the eighth decade and 6 in the ninth. There are 3 autopsies recorded in the eighth and 2 in the ninth. In all 3 of the eighth decade and one of the ninth, cerebral softening was found. One was due to thrombosis of the right internal carotid, secondary to embolism of the brachial artery. The other 3 appear to have been primarily thrombotic. The one case of cerebral hemorrhage occurred in a colored man, aged eighty-one, and was associated with intense degeneration of the whole vascular system.

**Symptoms.—Preceding.**—Slight cerebral symptoms may occur many months before apoplexy. These depend upon the vascular changes which lead up to the rupture or the occlusion of a vessel, and are important as indicating that such changes have taken place. It is obvious that an embolus whose origin is from some distant part of the circulatory system, can give rise to no cerebral manifestations until it lodges in an artery of the brain. But if the occluded artery be small, or one in which collateral circulation can be established, the symptoms may be slight or transient and premonitory of those following a subsequent larger embolus.

The liability of an artery to rupture depends upon some weakness of the vessel wall, usually upon the presence of small aneurisms. These dilatations on the smaller vessels themselves give rise to no symptoms that can be distinguished, and so in many, probably most cases of cerebral hemorrhage there are no premonitory symptoms. The associated cerebral arteriosclerosis may, however, cause symptoms for a long time before the occurrence of apoplexy, due to rupture or thrombosis.

The *rupture* of a larger aneurism of one of the arteries at the base of the brain is a rare cause of cerebral apoplexy. These dilatations usually give no symptoms until their rupture, but at times they cause the manifestations of cerebral tumors. When such symptoms are present, associated with subjective noises in the head, and the presence of a vascular murmur heard through the skull, the diagnosis of an aneurism is justified. This association appears to be so rare as to have very little practical importance. Much more frequently the aneurismal dilatations are associated with widespread arterial changes and we are apt to have the symptoms of cerebral arteriosclerosis. Hemorrhage may occur into the substance of a brain tumor, and so the symptoms of such a growth may at times be premonitory of cerebral hemorrhage.

The great majority of cases of cerebral thrombosis occur in the last half of life, and are due to more or less chronic changes of the cerebral

vessels—cerebral arteriosclerosis. The most significant symptoms due to this condition are a lowering of the mental strength, giddiness, slight cerebral attacks, convulsive seizures, transient aphasias, slight weakness of the various limbs, paresthesia and pain, etc., when associated with evidence of sclerosis in the peripheral arteries, especially in those of the retina, and the signs of general arteriosclerosis.

Thrombosis of a cerebral vessel, in the first half of life is most commonly due to the effects of syphilis, or is in association with acute infectious diseases. That due to syphilis may be associated with the symptoms which are recognized as indicating specific involvement of the brain, intense periodical headaches, paralysis of various cranial nerves, etc., and these we may in the present relation regard as premonitory. There are usually no premonitory symptoms of thrombosis of a cerebral vessel occurring in the course of acute infectious diseases.

The nervous symptoms which precede an apoplexy, if cerebral syphilis be excluded, are practically all due to arteriosclerosis. They more commonly precede thrombosis than hemorrhage, but are not exceptional even with the latter.

**Exciting Causes.**—An apoplectic attack may occur under all conditions of life—during sleep, while awake in bed, upon getting up, at table, at work, at play, or while occupied in any other manner. Overexertion does not often play its assigned important rôle in exciting the attack, and, indeed, it is rarely possible to discover from the history any evident relation of the occupation to the stroke.

In the analysis of the cases in which the onset was after the first decade, and excluding the traumatic cases, there were 379 in which the circumstances of the attack were definitely noted. Among these 125 occurred while the patients were in bed and in most instances asleep. Fifty-four of these occurred during the course of some other illness, and 37 while the patients were in the hospital. In 36 instances the attack was noticed when the patient got out of bed, and some of these may have occurred during sleep. In contrast to this it is interesting to note that but 4 cases occurred while patients were preparing for bed. Fifty-nine attacks developed while the patients were sitting. A number of these were while the patients were eating a meal, but in most instances they were quietly reading, sewing, etc. In 3 instances they were playing cards, and it is possible that emotional effects may have been operative here and in some other cases. This seemed certainly so in one instance, that of a colored midwife, who became very angry on account of the refusal of the authorities to admit her patient to the hospital. She fell unconscious, and was herself taken into the ward, and died within twenty-four hours. This was the only case in which anger seems to have played a part. Ten attacks occurred while the patients were on more or less protracted sprees, and 54 while the patients were simply walking about.

This analysis includes apoplexies due to all causes, and it might well be that those cases which took place during rest, included, for the most part, those due to thrombosis, and that the cases of hemorrhage were among those occurring with more or less bodily activity. Theoretically, anything that increases the blood-pressure within weakened vessel

walls would tend to cause their rupture, and we should expect to find many cases of apoplexy due to hemorrhage occurring during muscular exertion or those emotional states associated with increased action of the heart. Increased cardiac action might also be assumed to favor the detachment of an embolus, and so similar causes might be expected to be active in this class. Thrombosis, on the other hand, is favored theoretically by quite opposite circulatory conditions, and the predisposing causes, changes in the vessel walls or in the blood, being present, slowing of the blood current should act as an accessory factor, and might determine the time of the occlusion.

Clinically the state of affairs is not so simple, and is modified by many circumstances which cannot be determined, and it is very unsafe to assume the character of the lesion from the state of activity of the patient at the time of the stroke. The study of a large number of cases in which the lesions were determined after death would show the real relation of these factors. Unfortunately, the figures available from this series are but few, but they may help to swell the total from other sources.

There were 25 cases of *cerebral hemorrhage* in which the onset was given. Twelve of the attacks took place while the patients were at rest, 10 while in bed, and 2 while sitting quietly; 6 of the 12 attacks occurred in the hospital wards, as the terminal event of various conditions; 3 other apoplexies terminated alcoholic debauches; 1 of these patients was sitting at a table, and the others were probably not very active, although as to this we have no definite data; 4 patients were struck down while walking about, and 5 while at their every-day work. In only 1 case did there seem any great amount of physical exertion, and that was in a man, aged thirty-nine years, who, after a long bicycle ride, was found at 3 A.M. on the steps of a house, paralyzed and aphasic. The onset was noted in 23 cases of *cerebral softening*, and in 16 of these the patients were in bed, all but one being confined by some severe illness; 14 of them were in the hospital at the time of the onset. Of the other 7 attacks, 4 took place while the patients were at work, and one while occupied in each of the following ways—sitting quietly, walking about, and during an alcoholic spree.

It will be seen that of the 26 cases in which cerebral lesions developed while the patients were in bed, only 10 proved at autopsy to be hemorrhage, but that of the 19 apoplexies developing during the more or less bodily activity (including the 4 cases which occurred during alcoholic debauches), 13 proved to be due to cerebral hemorrhage.

A number of authors have interested themselves as to the importance of bodily activity in the production of the various cerebral vascular lesions. Ernest Jones<sup>1</sup> analyzed the records at University College Hospital, London, in relation, among other things, to the time of onset of the various vascular lesions. These records covered sixty-five years, and the available material consisted of 160 lesions, 123 cases of hemorrhage, 24 of thrombosis, and 13 of embolism. None of his cases occurred during excessive bodily activity. He compared his results with those of various

<sup>1</sup> *Brain*, 1905, xxviii, 527.

other authors, and concluded: "Rest in bed, and especially sleep, protect to some extent against cerebral hemorrhage. This would seem a more accurate statement than that rest and sleep predispose to thrombosis, as I can find no evidence in favor of this, although in actual practice the two statements have a similar bearing on diagnosis. Severe exertion and time of day appear to have had too much stress laid on them in the past. A military aneurism that is going to rupture rarely needs the aid of severe exertion for this consummation. Time of day is only of interest when the habits of the patient's blood-pressure at different hours are known."

**The Apoplectic Attack.**—Throughout the present article the word apoplexy has been used more in its original sense as meaning an acute cerebral attack, but it has been stretched to include not only those seizures of sudden loss of consciousness with paralysis of sense and motion, but also those in which the consciousness is affected later, or, indeed, not at all. Trousseau used the word strictly as signifying an absolutely sudden loss of consciousness, and so pointed out that "apoplexy" was rarely, if ever, the initial symptom in cerebral hemorrhage, and that if the patients were under observation at the time of the onset, symptoms due to cerebral lesions could always be noted before consciousness was lost. Indeed, he states that he had never seen a man struck down as by a blow and dropping instantly in a state of unconsciousness.

*Loss of consciousness* as the initial symptom is indeed rare, and in the vast majority of cases patients usually complain of headache, vertigo, tingling or weakness in some part of the body, as the indications of the attack. The loss of consciousness, if it occurs, may be delayed only a few minutes, or for several hours, or, indeed, days. The manner of occurrence of the different lesions—thrombosis, hemorrhage, and embolism—differs, and one would expect the development of the symptoms to be somewhat characteristic; thus, the closure of an artery by a thrombus is a more or less gradual process, and so one would expect the symptoms to develop more or less gradually. The rupture of a bloodvessel is a sudden occurrence, and the resulting symptoms should make their appearance quickly. The occlusion of a bloodvessel by an embolus may be absolutely sudden, and in connection with it we might expect apoplexy to develop in its classical manner.

These considerations are, in general, true, but are often so modified that conclusions based upon them cannot be accepted absolutely. Although it is true that thrombosis is not in its entirety a sudden process, still the final closure of a vessel may occur suddenly, and it is not at all uncommon to have the symptoms develop with great rapidity. Jones, in this connection, says, "There is a large group of thrombosis cases, the onset of which is apoplectiform." The percentage has been put as high as 58 by some authorities. There are a number of such cases in this series. For instance, a woman, aged fifty-two years, who had one previous attack from which she recovered, while in apparent health and transacting business with a lawyer, fell unconscious. After her death, three weeks later, cerebral thrombosis was found.

Although the rupture of a vessel is an instantaneous process, the tear at first may be very slight, and only a small quantity of blood may escape.



The focus may increase in size more or less gradually, destroying more and more of the nervous tissue, and many cases of cerebral hemorrhage have the onset typical of thrombosis. A man, aged fifty-nine years, while sitting with his family after supper, complained of intense headache. Ten minutes later it was noticed that his face jerked, and that when spoken to he did not answer questions. In half an hour he got up, closed the house, went up stairs, and moved about his room. He gradually became unconscious during the night, and was completely paralyzed on the right side. After his death, one week later, a large hemorrhagic clot was found in the left cerebral hemisphere. Another man, aged sixty-five years, with definite arteriosclerosis, gradually lost power on the left side in the course of twenty-four hours, with increasing dullness but no actual coma. He improved somewhat, but died in eleven days from bronchopneumonia. At autopsy the brain showed marked arteriosclerosis, multiple areas of softening, and, in the right hemisphere, a large clot, 7 cm. in diameter.

The stoppage of an embolus is always sudden, but if an embolus lodges at the bifurcation of a vessel, the blood current is not completely interrupted until the formation of the thrombus around the embolus occludes one or the other of the arterial branches. This secondary thrombus formation may extend and cut off branches that had previously received blood by collateral circulation. An embolus may be caught in an artery that has free collateral circulation, or there may be more than one embolus, the first plugging a relatively small vessel and a subsequent one a more important artery. It is obvious that if any of these circumstances occur the symptoms following an embolus of the cerebral vessels may develop more or less gradually.

**Consciousness.**—Even though it is true that an absolutely sudden loss of consciousness is uncommon in association with any of the cerebral vascular lesions, still consciousness is often disturbed with all of them. In the records of 401 apoplectic attacks in which the state of consciousness was noted, there was no loss of consciousness in 202 cases, but in 199 it was either lost or markedly disturbed. In 107 of these it was reasonably certain that the disturbance of consciousness occurred early in the attack. As but few came to autopsy they are of no value in determining the relative frequency of loss of consciousness in relation to the different lesions. Jones analyzed his own cases, together with those which he had been able to collect from the literature, and found that among 201 cases of embolus, consciousness was lost in 47.7 per cent., and was affected in nearly 60 per cent., while in 235 cases of cerebral hemorrhage consciousness was lost in nearly 75 per cent. He gave no collected figures for thrombosis in this relation, but in his own 22 cases there was loss of consciousness in 45.5 per cent.

The mechanism of the loss of consciousness is not thoroughly understood, but since consciousness depends upon an adequate supply of blood to the cerebral cortex, a disturbance of this is the most obvious cause for the loss. It is easy to understand how the sudden closure of one of the large vessels would produce such an anemia as to disturb consciousness, and, indeed, an embolus lodging in and occluding the middle

cerebral or the basilar artery, both favorite seats for such an occurrence, would offer the ideal conditions for a sudden loss of consciousness. Saveliew,<sup>1</sup> who analyzed all the cases of cerebral emboli in Virchow's laboratory from 1856 to 1893, states that a sudden loss of consciousness is the common onset in cerebral embolism. When the artery is slowly occluded there is more opportunity for collateral circulation; primary cortical anemia is less apt to occur, and consciousness when lost is apt to be lost late. This late loss of consciousness must be due to secondary causes, and in association with occlusion of a vessel is probably dependent upon the œdema which accompanies and surrounds the anemic area. This secondary œdema causes, among other things, an increase in intracranial pressure, which may be so great as of itself to produce a cortical anemia. Destruction of the brain tissue itself must have an effect on consciousness, for it depends upon the association of various cerebral activities, and must be affected by anything that injures the structure upon which these depend. We do not know just how great the disturbance must be to abolish consciousness or the relative importance in this relation of the different parts of the brain.

The loss of consciousness accompanying cerebral *hemorrhage*, although perhaps rarely the initial symptom, very commonly occurs early, and appears to be more frequent and usually of longer duration with this lesion than with either of the other two. The explanation is not clear, and many theories have been advanced. Gowers,<sup>2</sup> in speaking of the loss of consciousness, says: "It is probably first due to irritative inhibition of the cortex, which is maintained by increased intracranial pressure. The laceration causes powerful irritation, and when the hemorrhage is into structures that have a wide relation to the cortex, as in the pons, a small extravasation, which can have little effect on intracranial pressure, causes early loss of consciousness, which endures." von Monakow analyzed the various effects caused by a hemorrhage into the substance of the brain, and divided them into the following groups: (1) The primary local destruction of the nervous tissue; (2) the œdema in its neighborhood; (3) the decrease in the amount of blood in the vessels in the neighborhood of the clot; (4) accompanying circulatory changes in the cortex; (5) the transmission of the mechanical shock caused by the sudden escape of blood into the brain in all directions throughout the intracranial cavity, this shock, besides its physical action, acting as a physiological stimulus to various parts of the brain; especially, he thinks, it may stimulate the vasomotor centres and cause a reflex contraction of the cortical vessels; (6) increase in intracranial pressure; (7) interference with circulation of the cerebrospinal fluid. In the last analysis he believes that the disturbance of consciousness depends upon a sudden cerebral anemia, and that this is brought about by some or all of the various factors enumerated above, but that probably the mechanical stimulation of the vasomotor centres "plays the lion's part."

The occurrence and degree of coma does not depend upon the size of the hemorrhage, but seems to have some definite relation to its posi-

<sup>1</sup> *Virchows Arch.*, 1894, cxxxv, 112.

<sup>2</sup> *Brit. Med. Jour.*, London, 1907, ii, 1.

tion. The exact relationship has not been definitely determined, but von Monakow states that after analyzing a great number of cases it appears to him that coma is more apt to occur the nearer the lesion is to the central gray matter of the third ventricle, or the nuclei of the optic thalamus, and that it is much less apt to occur when the white matter of the cerebral hemisphere is involved. When the hemorrhage breaks through into the ventricle consciousness is almost always lost, and Jones found that it was lost in 85.5 per cent. of this class of cases, while in the non-ventricular hemorrhages the percentage of loss was only 69.4 per cent.

The *duration* of the coma varies greatly, and may be very short or persist until death. It is apt to be shorter when associated with embolism and longer with thrombosis and hemorrhage; but even in hemorrhage von Monakow gives the average duration as only from a half hour to four hours. Oppenheim puts the duration somewhat longer, from one to four hours for the lighter cases and one to two days for the more severe ones. From the actual coma, patients not infrequently pass into a state of dull consciousness, during which they have very little recollection of what passes around them, and it is not uncommon for them to insist that they were unconscious for a week or even longer.

**Pulse and Blood-pressure.**—During an attack of apoplexy the condition of the pulse varies markedly. A slow pulse, particularly with a high blood-pressure, indicates an increased intracranial pressure, and is usually associated with a cerebral hemorrhage. Thus, in an old colored woman, brought to the hospital directly after the attack, and who died within four hours from a large cerebral hemorrhage, the pulse on admission was only 48, and the blood-pressure was above 350 mm. Hg. In a man admitted on the third day after his stroke, the pulse rate was also 48, and the blood-pressure between 330 and 340 mm. Hg. A large capsular hemorrhage was found. These cases are exceptional and the rate of the pulse is often not much lowered, and is at times accelerated.

The character of the pulse is usually of more importance, for in association with cerebral hemorrhage it is, as a rule, hard and full, and the blood-pressure may be very high. Such an excessive elevation, although characteristic of acute increase in intracranial pressure, and perhaps never found clinically except with cerebral hemorrhage, is exceptional even with this condition. The blood-pressure more usually ranges between 150 and 200, and even in rapidly fatal cases may be quite normal as in a woman, aged forty-three years, brought to the hospital shortly after an attack, in whom the pulse was 100 and the blood-pressure 125 mm. Hg. She died within twenty-four hours, and a large hemorrhage was found in the right hemisphere.

A blood pressure in the neighborhood of 200 is frequently found in association with arteriosclerosis, and as this condition often precedes for a long time the occurrence of an apoplexy due to either rupture or thrombosis of a cerebral vessel, the significance of a high blood-pressure during an apoplectic attack depends largely upon our knowledge of the previous conditions. Although it is generally stated that when an apoplexy has been due to thrombosis the pulse is quick, intermittent, and of low tension, it is not at all uncommon to find a high blood-pressure

associated with this condition. Janeway<sup>1</sup> says: "Thrombotic softening is the common cause of hemiplegia in our patients, even when they have granular kidneys and high blood-pressure."

Changes in blood-pressure are more significant than the actual height, and a rapidly rising pressure indicates an increasing cerebral compression, while a falling pressure is one of the most definite signs of failure of the vasomotor centres and of approaching death. A good illustration of the first condition is the case of a man brought into the ward shortly after having had a capsular hemorrhage. His blood-pressure was then 220 mm. Hg. and the pulse about 90. The pressure rose steadily, and the next day reached 250, when an operation was undertaken. As the dura was opened the blood-pressure fell to 180 and subsequently even lower. It arose again, but never reached 200 during the three days which he lived. A most impressive example of the second sequence of events was in a man whom we had observed for a number of months. He showed the typical picture of cerebral arteriosclerosis, and had suffered from several vascular attacks. He was in the ward for nearly a month before the final accident. During the first few days of his stay in the hospital his blood-pressure ranged between 230 and 240, but subsequently about 215, with a pulse rate of 95. One afternoon he complained of intense headache, for the relief of which morphin had to be given. At 6 P.M. the blood-pressure was 200. In the middle of the night he was found in coma. His blood-pressure at 3.30 A.M. was 240; pulse, 145. At 5.30 the pressure was 200; at 11 it was 180, and then fell quickly, reaching 80 at 1 P.M. and 60 at 1.15 P.M., the pulse remaining between 120 and 130. Death took place at 1.40 P.M. At autopsy a large cerebellar hemorrhage was found.

**Respiration.**—The respiration is usually slow and deep; it is often stertorous and irregular, and at times shows the Cheyne-Stokes type. The noisy stertorous breathing is usually due to the fact that during coma the mouth has a tendency to fall open and the tongue to fall back. Bowles<sup>2</sup> points out the importance of keeping patient on his side.

Cheyne-Stokes breathing during apoplexy is usually a symptom of the embarrassment of the respiratory centres in the medulla as the result of anemia, secondary to increased intracranial pressure, and is an important danger signal. The vasomotor centres tend to keep the blood-pressure above intracranial pressure, so that the medullary centres may still be supplied with blood. When the intracranial pressure remains constantly high, the blood-pressure may show periodic waves (Traube-Hering waves), varying from a little below to a little above the level of the intracranial pressure. Experimentally, when such variations occur, respiration becomes intermittent. In general, it ceases when the blood-pressure is relatively low and begins and increases when the blood-pressure rises above intracranial pressure. If careful tracings of the blood-pressure and respiration be made in a case of apoplexy showing the Cheyne-Stokes respiration, quite analogous conditions will be seen to be present. Eyster,<sup>3</sup>

<sup>1</sup> *The Clinical Study of Blood-pressure*, 1907, p. 189.

<sup>2</sup> *On Stertor, Apoplexy, and the Management of the Apoplectic State*, 1891.

<sup>3</sup> *Jour. of Exper. Med.*, 1906, viii, No. 5.

from a study of such cases, concludes that this type of Cheyne-Stokes breathing is due to the centre of respiration being intermittently supplied with blood; for when the centre has been deprived of arterial blood, it ceases to be capable of responding to its normal stimulus, *i. e.*, an increase in its supply of carbon dioxide and a reduction in oxygen, and so when the arterial pressure falls below intracranial pressure the medulla becomes anemic and respiration ceases. Anemia of the medulla excites the vasomotor centres to renewed activity, blood-pressure is again raised, circulation is reëstablished through the medulla, the respiratory centre regains its irritability, responds to stimuli, and respiration commences. The respiratory efforts vary in force in accordance with the varying relations between the irritability of the centre and the force of the stimulus that excites it.

There are often other phenomena due to the varying blood supply of the brain. The pulse varies in rate, being slow during the period of apnœa, as a result of the stimulating effect which anemia has on the cardio-inhibitory (vagus) centre. This inhibition is relaxed when respiration begins and the pulse rate is increased during the period of inhibitory activity. The mental activity also varies, and the patient, who is completely unconscious when the respiration ceases, can be aroused and may speak voluntarily, during the period of respiratory activity. At times the pupils show a periodical change in their size, dilating at the beginning of apnœa and contracting when respiration returns (Cushing). The importance of this type of periodic respiration, especially when accompanied by these other phenomena, is as an indication of an increase of intracranial pressure to such a degree that the vasomotor centre has difficulty in maintaining the blood-pressure above the intracranial pressure, and is a symptom of the utmost gravity.

**Ophthalmoscopic Examination.**—This may give important information, both as to the state of the bloodvessels and as indicating the condition of intracranial tension. Bordley, who examined many of the cases in the hospital, states that in most cases no other fundus changes were seen except those which preceded the stroke, *i. e.*, the changes indicating degeneration of the bloodvessels. In certain patients, however, particularly in those who were profoundly unconscious and showed Cheyne-Stokes respiration, undoubted choked disk was present. This was often of low grade, and with more dilatation and tortuosity of the veins than is usually seen with such a degree of choked disk, but it was seldom associated with retinal hemorrhages and exudates. In the cases of traumatic intracerebral hemorrhage, in which there was rapid increase of intracranial pressure, choked disk was constant.

**Temperature.**—When an apoplexy is due to cerebral hemorrhage there is usually an initial fall of from  $1^{\circ}$  to  $2^{\circ}$ . This may last only an hour or two, to be followed by a gradual rise, and may be entirely missed unless the patient is under observation at the time of the stroke. It is said that with thrombosis this initial fall does not take place. The writer has not sufficient exact data of cases in which the lesion was proved at autopsy to express any definite opinion. The cases of thrombosis which occurred in the hospital were in sick patients who usually had more or less fever

and the changes in the temperature at the time of the stroke were neither constant nor remarkable.

The temperature almost always rises within the first twenty-four hours, and in rapidly fatal cases may reach great heights. Gowers associated an immediate elevation of temperature with hemorrhages into the pons or medulla. This hyperpyrexia is of great significance, and whenever it occurs is usually a sign of approaching death. In a patient already referred to, who died in about twelve hours from a cerebellar hemorrhage, the temperature rose rapidly to 106.5°. The attack took place in the night, and the initial fall of temperature, if it occurred, was not determined. On the other hand, in certain quickly fatal cases the temperature continues to fall from the time of the attack, and may reach remarkably low levels. A rectal temperature of 95° has been recorded. The cause of these extreme temperatures seems to be a disarrangement of the heat regulating mechanism, and is one of the signs of failure of the cerebral functions.

The ordinary rise of from 1° to 2° in the first few days after an apoplexy has no especially grave significance. It is the so-called fever of reaction, and is believed to depend upon some inflammatory process about the hemorrhagic focus. This fever usually lasts for only two or three days, after which the temperature remains normal. Should the fever be higher than usual or last longer, it becomes of grave significance, and at any time if the temperature rises rapidly to unusual heights the prognosis becomes very grave.

**Signs of Local Injury to the Brain.**—The symptoms so far considered are general, but even during the height of an apoplectic stroke there are in most cases signs of local injury. These depend somewhat upon the location of the lesion, but for the most part indicate that the functions of one cerebral hemisphere have been disturbed. These symptoms are usually paralytic but may be irritative, when they are revealed by an excess of muscular activity. The limbs on the side of the body opposite to the lesion may be in a state of hypertension (early rigidity) with the eyes and head deflected toward the side of the spastic limbs and away from the lesion—*spastic conjugate deviation*; or the affected limbs may be the seat of clonic convulsions, and if the lesion is in the motor cortex, or near it, typical Jacksonian epileptic attacks may follow.

Symptoms of *paralysis* are much more common, and all the movements represented on one side of the brain are usually more or less affected. During coma, when the patient is making no spontaneous movements, this is often shown by a decrease in the muscle tone on the side opposite to the lesion. The limbs are flaccid, they fall like a dead weight when raised and released, while those on the opposite side may show a certain amount of resistance. Heilbronner<sup>1</sup> points out that this lack of muscular tone may be determined by inspection, for if the patient be on a firm support the muscle mass on the affected side, particularly that of the thigh, acts like a sac filled with a semi-solid, and takes the shape determined by gravity. It is broadened and flattened, while that on the normal

<sup>1</sup> *Deutsche Ztschr. f. Nervenhe.*, 1905, p. 1.

side has a more rounded contour. The lack of tone of the muscles of the face on the affected side is shown by the cheek being puffed out more than on the other side during forcible respiration, the air escaping from the corner of the paralyzed side of the mouth. At times the nostril on that side, instead of being expanded, is contracted during inspiration.

Not infrequently the patient exhibits a tendency to lie with the head turned toward the non-paralyzed side and with the eyes deflected strongly toward that side, *i. e.*, the patient looks toward his lesion. This is *paralytic conjugate deviation*, and is due to the fact that in each cerebral hemisphere movements are represented which turn the eyes and head to the opposite side and a destructive lesion paralyzes them. The normal position of the eyes depends upon the delicately adjusted activity of their muscles, and so when the tone of the muscles associated in a definite movement is decreased, the tone of the muscles causing the movement in the opposite direction deflects the eyes. Thus a lesion in the right hemisphere causes a decrease of the activity of the muscles which turn the eyes to the left, *i. e.*, the left external rectus and the right internal rectus, together with the muscles active in the closely associated movement of the head to the same side, and it is the unbalanced tone of the muscles which opposes these movements which causes the deflection of the head and eyes to the opposite side.

Muscle tone depends, for the most part, upon reflex stimulation, and it seems probable that conjugate deviation in apoplexy is, to a large extent, due to a disturbance of the reflex mechanism. At any rate, the symptom is usually transient, occurring during coma, and lasting from a few hours to one or more days, and is coincident with disturbances of other reflexes. When it persists for a longer time it is often associated with hemianopia, a condition in which all the visual stimuli come from but one-half of the patient's surroundings and tend to keep the eyes turned toward that side. The mechanism is a complicated one, and many stimuli must be active in maintaining the tone of the centres. That the visual impressions, although normally perhaps the most important, are not the only ones concerned, is shown by the interesting case reported by Dejerine and Roussy,<sup>1</sup> in which conjugate deviation of the head and eyes occurred in an old woman who had been blind since infancy.

**Pupils.**—During the apoplectic attack these are usually equal and about normal size. They may be dilated, contracted, or unequal, with the larger pupil on the side of the lesion when this is in a cerebral hemisphere, but on the opposite side when the focus is in the brain stem. During coma the light reflex is absent.

**Reflexes.**—During absolute coma all reflexes are abolished, but very generally the coma is not of that grade, and one often is able to demonstrate a difference in the reflex activity of the two sides. In general, it may be stated that a sudden severe vascular lesion in one cerebral hemisphere has a tendency to abolish the reflexes on the opposite side of the body. The early conjugate deviation of the eyes and the lack of

<sup>1</sup> *Rev. Neurol.*, 1905, xiii, 161; Weisenburg, *Jour. Am. Med. Assn.*, 1907, xlviii, and Debray, *Jour. de Neurol.*, October 5 and 7, 1907, have given interesting reviews concerning this subject.

muscle tone on the paralyzed side must, in a large measure, be due to this effect. The reflexes from the skin and mucous membranes, as well as the deep reflexes, may also be lost or altered, and when such a change is demonstrated on one side it is an important indication of a unilateral cerebral lesion. Since Babinski pointed out the varying response of the great toe to plantar stimulation under different conditions, this reflex has assumed the utmost importance. If a lesion has occurred which implicates the pyramidal tract, the great toe, instead of being turned down, moves in the opposite direction, *i. e.*, in extension, or more properly dorsal flexion. This abnormal response (Babinski reflex) may appear directly after a cerebral lesion, and may be the first positive indication that such a lesion has occurred. The writer has noted this on several occasions, and in a patient whose attack occurred in the ward it was marked within twenty minutes.

As the coma lessens the reflexes return, first on the non-paralyzed side, and according to von Monakow in the following order: first the conjunctival reflex, then the pupillary light reflex, the abdominal and the other skin reflexes, and finally the deep reflexes. The return of the reflexes on the paralyzed side may be delayed for a considerable time, even, it is stated, for two or three weeks. In our experience, however, the deep reflexes return quickly, and may even be exaggerated within a short time, although the corneal and the abdominal reflexes may remain absent. The importance of the absence of these two skin reflexes in association with hemiplegia has been insisted upon.<sup>1</sup>

During coma the bladder and rectum are apt to empty involuntarily, but there may be retention, and as the patient recovers there is not infrequently some difficulty in completely emptying the bladder.

**Paralysis of Voluntary Motion.**—Unless the coma is absolute, observation, in the great majority of cases, reveals that the patient has lost certain movements. It will be noted that one arm and one leg will lie perfectly quiet, while the limbs on the other side may be moved restlessly about; that when the patient is moved in bed or irritated in any other manner, the movements which result are only one side of the body. This lack of motion may also affect the face on the same side, *i. e.*, the patient has hemiplegia. At first, and in severe cases, all the unilateral movements are affected, but certain movements show a tendency to recover quickly or to be spared when the process is less intense. This selective paralysis in hemiplegia is of great interest and will be discussed in a subsequent section. The limbs on the non-paralyzed side are not infrequently moved restlessly about; the patient picks at the bedclothing, moves the hand to the head, draws up the leg, and may persist in these movements so constantly that at times they may suggest convulsive phenomena.

The movements of the chest during respiration are generally recorded in our histories as being equal and we have endeavored to confirm the common statement that the movements on the paralyzed side are less than on the opposite side.

<sup>1</sup> Milian, *Le Progrès méd.*, May, 1909.



**Diagnosis.**—The diagnosis of coma due to a cerebral vascular lesion is usually made without difficulty, as when a patient who has shown evidences of the circulatory conditions which predispose to rupture or occlusion of an artery has symptoms of local involvement of the brain (numbness and weakness on one side of the body, unilateral muscular twitchings, etc.) and then passes into unconsciousness, during which definite evidences of hemiplegia are present. At times the diagnosis may be difficult, and, indeed, impossible, as when the coma occurs suddenly without premonitory cerebral symptoms, or when the physician sees the patient for the first time during the attack, the onset of which has been unobserved.

*Coma* may occur under a variety of circumstances, and is the frequent terminal state of all fatal illnesses. It is often an evidence of the toxemias associated with nephritis (uremia) and diabetes. It occurs as a result of poisoning by alcohol, opium, and its derivatives, illuminating gas (carbon monoxide), chloroform, ether, etc. It may be associated with sudden disturbances of the general circulation, if they are sufficient to produce cerebral anemia, as in the ordinary fainting fit, the attacks of unconsciousness with Stokes-Adams disease, the rupture of a large aneurism, etc. It follows an epileptic attack and may be purely hysterical. It happens occasionally in malaria and heat stroke, and it is a frequent result of trauma.

In most cases the history will exclude many of the conditions in which coma may occur, and the mistakes are apt to happen when little or nothing is known of the patient or of the onset of the attack, as in hospital and police practice. Every such case should be regarded with suspicion, and the diagnosis of one of the less serious states should be deferred until the indications are clear and unmistakable.

When the coma is so deep that all special activities of the brain are in abeyance, when no movements occur and no reflexes can be elicited on either side of the body, evidences of a focal lesion of the brain are, of course, absent, and as it is upon these that the diagnosis of a cerebral lesion must largely depend, it may for a time be quite impossible to determine the cause of the coma.

*Uremia.*—In the absence of unilateral cerebral symptoms the diagnosis must depend upon the previous history, the occurrence of chronic nephritis, convulsions ushering in the coma, which usually develops rather slowly, etc. Objectively, the condition may be remarkably similar to apoplectic coma. The high arterial tension, Cheyne-Stokes respiration, and the ophthalmoscopic picture, may be quite similar in both conditions, and when it is remembered that transient focal cerebral symptoms are not uncommon in uremia, and that apoplexy frequently occurs in patients suffering from chronic nephritis, it will be seen how complicated the diagnosis may become. Eyster pointed out that in the periodic respiration of uremia the relation between the respiratory phases and the changes in the blood-pressure differs from that in Cheyne-Stokes respiration secondary to increased intracranial pressure, as in increasing cerebral hemorrhage, and it is probable that careful tracings of the respiratory and blood-pressure changes may give important indications. Simple

choked disk may be present in both conditions, but in nephritis the characteristic picture of albuminuric retinitis is much more frequent.

Coma occurring in pregnant women at about the time of labor is usually due to toxemia (uremia or eclampsia), but exceptionally it depends upon a cerebral hemorrhage, which, unless accompanied by focal symptoms, is very likely to be overlooked.

*Diabetic coma* generally causes but little confusion. The history of the preceding illness is usually obtained. The coma develops gradually, being preceded by lassitude, headache, great restlessness, and other symptoms. The pulse is apt to be rapid and of low tension, and the respirations show "air hunger," being full and noisy, but quite regular and usually not increased in frequency. The breath has a fruity odor, and so may the urine, which shows the presence of sugar, diacetic acid and acetone. Very rarely optic neuritis is present.

It must be remembered that the deep reflexes are often lost in diabetes, and their absence should not be given undue significance. Confusion may also arise from the fact that hemiplegia, in which no anatomical lesions were found, has been noted in diabetes. On the other hand, sugar has been found in the urine as the result of a cerebral vascular lesion. It is always safer to assume a definite focus when the symptoms indicate its presence.

*Alcoholic Coma.*—The mistake of regarding a case of apoplexy as one of acute alcoholic intoxication has been made innumerable times, and whenever this question arises the patient must be given the benefit of every doubt. A history of former alcoholism, and a marked alcoholic odor of the breath and vomitus have significance, but it must be remembered that alcohol is universally administered as a medicine under all conditions, and may have been swallowed at the onset of the attack, and also that apoplexy not infrequently occurs during an alcoholic debauch. The coma due to alcohol is almost never absolute, and the patient can usually be aroused to show some evidence of activity. This fact permits the discovery of paralytic symptoms, and it is the absence of these in such a patient, with the history, that justifies the probable diagnosis of drunkenness.

*Opium Poisoning.*—In acute opium poisoning the coma simulates intense drowsiness; the patient can be aroused by energetic stimulation, but lapses again into unconsciousness as soon as he is undisturbed. The pupils are contracted to an extreme degree; the respirations are stertorous, very slow, and may be regular, but at times occur in groups, as in the Cheyne-Stokes type. The skin is cold and of a livid hue; the pulse is rapid and of small volume. This picture is so characteristic that even without the history of administration of the poison mistakes are not apt to occur. It must not be forgotten that very small pupils sometimes occur with a hemorrhage into the pons or mid-brain, and that they may be associated with some preëxisting condition, such as tabes and general paralysis, in which apoplexy is not uncommon.

Poisoning by other agents rarely gives rise to much confusion, but where nothing is known of the history of the patient one should be on the alert not to overlook them. The burning of the mouth and the intense

gastro-intestinal disturbances, associated with symptoms of collapse, are usually sufficient to indicate the presence of carbohic acid poisoning. In unconsciousness from carbon-monoxide poisoning the history is almost always obtained. In doubtful cases the blood must be examined. Its bright cherry-red color and the characteristic reactions of carbon monoxide are conclusive.

Coma following changes in the *general circulation* is clearly distinguished by the associated conditions, and rarely gives rise to confusion. The ordinary fainting attack (syncope) is so well known as to require no comment. The attacks of unconsciousness in heart block are transient, and are so definitely associated with a pause in the ventricular pulse that mistakes should not occur. Rapidly occurring coma from the rupture of an aneurism, which persists until death, may, at times, be confused with that due to a large cerebral hemorrhage. The history and physical signs are usually sufficient to indicate the presence of an aneurism, and death is, in such cases, more rapid than in even the most severe apoplexies.

Absolutely *sudden death*, which is so often recorded in death certificates as due to apoplexy, is usually associated with a stoppage of the heart due to an obliteration of one of its coronary arteries. Apoplexy, even in the most quickly fatal cases, lasts a considerable time. Death within one or two hours is a rare occurrence, and the case recorded by Abercrombie nearly one hundred years ago, in which death occurred in five minutes, has not so far as the writer knows, been equalled. The quickest death in our records occurred in a little less than an hour, and in this case, besides the cerebral hemorrhage, there was a ruptured mesenteric aneurism.

*Epilepsy and Hysteria.*—The stupor following an ordinary isolated epileptic attack is usually easily recognized. The history of previous attacks, the characteristic convulsion which immediately preceded the coma, and its relatively short duration are significant. A case seen for the first time in the status epilepticus might easily be mistaken for one of convulsions and coma, following a cerebral vascular lesion, and the diagnosis would have to depend upon the subsequent course or the results of an autopsy. Unilateral epileptic convulsions are often followed by evidences of transient paralysis—a slight hemiplegia—even when there is no gross cerebral lesion; and, on the other hand, in patients past middle life, Jacksonian epilepsy is not infrequently the result of a previous cortical vascular lesion; and, indeed, such attacks may be the signs of a cerebral arteriosclerosis and the premonitory symptoms of a gross vascular lesion. In general paresis convulsive seizures and transient paralytic phenomena are not uncommon, but as cortical lesions often occur in this disease, one must be cautious in estimating the importance of an individual attack.

*Hysteria* at times introduces most puzzling and interesting questions of diagnosis. It should always be remembered, what is so often forgotten, that a hysterical patient has the same liability to organic disease that any other similarly situated individual has, and that in a predisposed individual hysterical symptoms may be so added to those which are dependent upon an organic lesion as to seriously confuse the picture. The definite

evidence of organic disease has the same significance in a hysterical patient as in any other condition.

Hysterical coma has the appearance of deep, normal sleep, which is unduly prolonged or which has occurred during the daytime after some psychical shock. The patient can usually be aroused, at least for a time, by painful stimuli, as a faradic brush, pressure over the supra-orbital nerve or the eyeballs, etc., or by the determined command of the physician or the suggestion of some disagreeable procedure. The pulse and respiration are usually not markedly altered, although at times the respiration shows most bizarre changes with entire absence of regularity. It may have an extremely rapid rate, suggesting the panting of an animal, or it may be so infrequent as to give rise to the fear that the patient is about to die from suffocation; and in very rare instances it shows the periodicity of the Cheyne-Stokes type. Persistent hiccoughing may be an alarming and confusing symptom, as this is also a symptom of an organic brain lesion. The temperature, unless in exceptional cases, is quite normal.

When such a condition has been ushered in by a typical hysterical convulsion one may feel reasonably sure of the diagnosis. The convulsions, however, may be absent or far from typical, and the condition so obscure that it is only after a detailed and fruitless search for some evidence of an organic lesion that a tentative diagnosis of hysterical coma is justified. The diagnosis of hysteria, especially where symptoms, apparently grave, are present, should never be more than a provisional one, and should stimulate to repeated examinations. The paralyses associated with hysterical coma are to be distinguished by the absence of the distinctive characteristics of an organic type of paralysis.

*Malaria and Sunstroke.*—The sudden loss of consciousness that occurs exceptionally in malaria, and, as a rule, in heat stroke, may simulate the coma of apoplexy, and in malarial districts the diagnosis is often difficult. High fever is common in both malaria and heat stroke, and in the latter it may reach a very high point, 108° or even more. The discovery of the malarial parasite in the blood is conclusive of that disease, and the blood should always be examined in cases of coma where there is a possibility of malaria being the cause. The diagnosis of sunstroke depends upon the history of the onset under conditions that may induce it, and the absence of the evidences of paralyses characteristic of apoplexy.

*Trauma.*—In coma following injury to the skull the history of the blow is usually clear, but in a case in which the history is unknown, or the circumstances arouse suspicion of foul play, a careful examination of the head must be made. Many of the symptoms following injury to the head are due to hemorrhage within the cranium and give rise to the same localizing symptoms found in spontaneous apoplexy. It is in these cases of hemorrhage, usually from a meningeal vessel, that the symptoms of an increasing intracranial pressure are most characteristically shown. Such evidences, however, do not always mean an increasing hemorrhage, for they may be due to œdema which follows the trauma.

From what has been said it follows that it is often difficult to determine at once whether coma is due to a cerebral vascular lesion or some other cause, but if the inquiry be made carefully and in such a manner as to

reveal the presence of the various conditions most likely to cause mistakes, and the conclusion not too hastily made, grave errors can usually be avoided. The history must be gone into carefully and confirmed in every way possible, the patient examined for the evidence of any bodily injury, the urine obtained and examined, the bodily temperature, respirations and pulse watched carefully, the blood examined, and the ophthalmoscopic examination must not be omitted.

Important as these procedures are, the proof that there has been a local injury to the brain and that the coma is associated in some way with the cause of the lesion, depends upon the discovery of some irregularity in the nervous activities pointing to such a lesion. These symptoms have been considered but it is at this stage in the examination that the necessity for a systematic search for them must be kept in mind.

The attitude of the patient must be observed. Is he quiet or restless? If restless, are the movements confined completely or largely to one side of the body? Are the movements purposeful or not? If convulsions occur, it is of the utmost importance to note the movements which are first involved; their march, extent, and duration. Is there any difference in the muscle tone of the two sides of the body? If there is such a difference, are the limbs abnormally flaccid or unusually stiff?

Does the patient tend to take any particular attitude? Is the head turned to one side and held there? Are the eyes in the median line or turned to one or the other side? Are they steady, or do they move and to what extent? What are the conditions of the pupils as to their size and reaction to light? Is there any asymmetry of the face during respiration? Do the two nostrils dilate equally on inspiration? Is one cheek blown out more than the other during expiration? Are the visual apertures equally wide? Are the wrinkles on one side of the face more pronounced than on the other?

The movements of the chest and abdomen during respiration should be carefully watched to see whether there is any difference between the two sides, and this should be observed both in quiet and forced breathing.

Does the patient respond to stimuli upon the two sides of the body equally, *i. e.*, is there any evidence of disturbed sensation? Care must be taken not to mistake simple reflex responses for the more elaborate movements that we believe to depend upon the activity of the cerebral cortex.

If the patient is sufficiently conscious, tests should be made to determine whether he sees objects on both sides, so as to exclude hemianopia.

The reflexes must be carefully investigated, particularly as to the relative activity on the two sides. The reflex movements that follow stimulation of the cornea, the nasal mucous membrane, the skin of the abdomen, and, in man, of the inner side of the thigh, are the most important of the so-called skin or superficial reflexes. Babinski's reflex and its modifications must be tested for with great care. The deep reflexes must be noted.

The condition of the skin of the patient is important. Is there any difference in the two sides as to the color of the skin, its temperature, and the amount of visible perspiration upon it?

When unilateral symptoms are demonstrated the presence of a local

lesion of the brain is strongly suggested, and when the symptoms have developed in a sudden apoplectiform manner the diagnosis of some one of the vascular lesions is usually justified. At times, however, confusion may be caused by *hysterical paralysis*. In this there is often complete paralysis of the limbs on one side of the body, with marked disturbances of sensation, which may have developed suddenly after a period of real or apparent unconsciousness. At times, even with a thorough understanding of hysteria, together with a knowledge of the characteristics of organic paralysis, the diagnosis may be extremely difficult. Usually, however, the hysterical nature of the paralysis is manifest. The attack occurs in a hysterical subject after some psychological shock. It usually develops somewhat slowly and without loss of consciousness. The loss of motion is confined, in the great majority of cases, to the arm and leg, and spares the face and the other muscles supplied by the cerebral nerves. In the limbs affected all movements are completely lost, and the paralysis does not show the selective distribution which follows a cerebral lesion. The paralyzed limbs are often quite flaccid, but contractures do not infrequently occur. There is very generally a loss of cutaneous sensation which is apt to involve the whole affected side, including the head. The anesthesia also involves the mucous membranes, and may include the nerves of special sense.

In sharp contrast to this absolute loss of function, depending, as we believe, upon some abnormality in the field of consciousness, is the quite normal activity of the lower nervous centres. There is no disturbance in the functions of the bladder and rectum, and the pupils react normally to light. The deep reflexes, although at times very active, are not so exaggerated as to cause a clonus, and are usually equal on the two sides of the body. This is also true of the reflexes from the skin. Plantar stimulation gives the normal plantar flexion of the great toe.

The hysterical loss of function is not infrequently influenced by suggestion. This is particularly true of the anesthesia which at times can be dispelled or transferred to the opposite side by various procedures. The paralysis may be similarly influenced and absolutely disappear after tetanization of the muscles by the application of the faradic current to the nerve trunks.

Under normal conditions the muscular activity involved during any strong voluntary effort spreads far beyond the muscles primarily concerned, *i. e.*, there is a synergistic action of many other muscles. This may consist of contraction or relaxation, and may even involve movements of the opposite side of the body. It seems to depend upon an automatic action of the lower centres,<sup>1</sup> and should not be affected in hysteria. Hoover's<sup>2</sup> observation seems to be an illustration of this. He has shown that when a normal person lying on his back endeavors to lift the extended leg from the couch, the opposite leg is pressed downward, and that in an organic hemiplegia this same pressure occurs when the patient tries to lift the paralyzed leg. If in such a case the patient lifts the unparalyzed leg the downward pressure of the paralyzed leg is

<sup>1</sup> Sherrington, *Quart. Jour. Exper. Phys.*, 1909, ii, 109.

<sup>2</sup> *Jour. Am. Med. Assn.*, 1908, li.

proportionate to its remaining strength. If the hemiplegia is due to hysteria the action is different. When the patient raises the non-affected leg the opposite leg, totally paralyzed for voluntary effort, is pressed strongly into the couch, while if he be told to endeavor to raise the paralyzed leg there is no movement on the non-paralyzed side, indicating that no outgoing stimuli have left the cerebral cortex, just as if he were consciously simulating the paralysis. Risien Russell<sup>1</sup> called attention to a somewhat similar phenomenon in hysteria, but in this case it concerns the lack of relaxation in the antagonistic muscles.

Hysterical hemiplegia was formerly believed to affect the left side much more frequently than the right, but Ernest Jones,<sup>2</sup> in an analysis of 277 cases, found no marked difference between the two sides; indeed, the right side was a little more frequently affected than the left, the percentage being 54.2 on the right side and 45.8 on the left.

It is often stated that hysteria may simulate every type or form of organic paralysis, but this simulation is usually superficial and disappears before a detailed examination. Mistakes are more commonly made by overlooking what is at least equally true, that many organic diseases may simulate hysteria, and that it is not at all uncommon to have a combination of the two conditions. This was well illustrated in the case of an unmarried woman, aged forty-five years, who after a long and severe emotional shock had an attack of dizziness associated with paralysis of the left arm and leg. She was admitted to the hospital in a highly nervous state, and the diagnosis of hysteria was made. The paralysis of the arm cleared up rapidly under suggestion, but that in the leg developed the typical signs of an organic paralysis. The distribution was characteristic, as was the spastic condition with well-marked ankle clonus. The patient was subsequently under my care for a number of years, and passed through numerous hysterical storms, but the paralysis of the leg remained constant and resisted all forms of suggestive treatment.

**Diagnosis of the Vascular Lesion.**—After the diagnosis of apoplexy has been made, *i. e.*, that the patient is suffering from the effects of a sudden vascular lesion, it becomes of great importance to determine, if possible, whether there has been a rupture of an artery and blood has escaped into the brain, or whether an artery has become plugged by a thrombus or by an embolus, causing cerebral softening. The diagnosis of the cause of the apoplexy in any individual case can be made only with a certain degree of probability. This degree may be so high as to almost reach certainty, or, on the other hand, so low as to amount to practically little more than a guess. Unfortunately, in the majority of cases the diagnosis has to remain very uncertain.

The brain lesion is the result of a vascular state and it is this that must be kept in mind. The vascular conditions liable to occur at the various periods of life differ, and so the age of the patient is an important point in diagnosis. In childhood and youth the spontaneous rupture of a cerebral vessel is rare, and apoplexies at that time of life, when not dependent upon inflammation, are very generally due to cerebral softening

<sup>1</sup> *Brit. Med. Jour.*, 1908, i, 608.

<sup>2</sup> *Rev. Neurol.*, 1908, p. 193.

and are in association with some definite cause, acute diseases, syphilis, etc., for thrombosis, and endocarditis for embolism.

The diagnosis of cerebral *embolism* depends upon the determination of the source of the embolus, and when such is found the inference is usually correct. Gowers points out that the presence of endocarditis is not excluded by normal heart sounds, and under conditions in which it is common, chorea, for instance, it should be suspected even when it cannot be determined. In the later decades, when degenerative changes in the vessels have become common, the association between valvular heart disease and apoplexy has less significance as indicating embolism, but even in early life one is occasionally surprised to find at autopsy a rupture of a vessel when its occlusion was confidently expected.

Between twenty and forty most cases of apoplexy are due to *thrombosis* secondary to syphilitic arterial disease, and then, or even before and after this period, when the cerebral accident occurs within a few years of the primary syphilitic infection, thrombosis is the most probable.

Cerebral *hemorrhage*, in spite of its rarity in the first half of life, should be suspected whenever the apoplexy has the distinctive characteristic of such an accident, especially when it occurs in apparent health in a subject in whom syphilis is unlikely or in conditions in which hemorrhages are common, leukemia, pernicious anemia, eclampsia, etc.

In the latter or degenerative period any one of the vascular conditions may occur. It would be important to know their relative frequency, but accurate knowledge could be obtained only from mortality statistics in large communities in which the cause of death was determined by autopsy.

In the cases dying in the Johns Hopkins Hospital softening was more common than hemorrhage, there being 30 of the former and 26 of the latter. Table VI shows the occurrence of these lesions in the various decades at the time of death. Ludlum,<sup>1</sup> who analyzed the material in Spiller's laboratory found a still greater frequency of softening, his figures being 69 of such lesions to 24 of hemorrhage; but Cadwalader,<sup>2</sup> using the same material and also that of the Pennsylvania Hospital, collected 72 cases of hemorrhage and 50 of softening. In the Royal Victoria Hospital, Montreal, the figures of which Dr. Adami put at my disposal, the relative frequency of the lesions was about that found at the Johns Hopkins Hospital, being 35 of softening and 29 of hemorrhage. These and other figures are given in Table VII.

TABLE VI.—*Johns Hopkins Hospital.—Fatal Cases.*

	Decades.									Total.
	1	2	3	4	5	6	7	8	9	
Cerebral softening	1	0	3	3	8	9	2	3	1	30
Hemorrhage . .	0	0	1	4	4	12	4	0	1	26

<sup>1</sup> *Jour. Nerv. and Ment. Dis.*, 1909.

<sup>2</sup> *Jour. Am. Med. Assn.*, 1914, lxii, 1385.



TABLE VII.

	Hemorrhage.	Softening.
Johns Hopkins Hospital . . . . .	26	30
Dr. Ludlum (Philadelphia Hospitals) . . . . .	24	69
Royal Victoria Hospital, Montreal . . . . .	29	35
Montreal General Hospital, Dr. Duval . . . . .	53	44
Boston City Hospital, Dr. Mallory . . . . .	132	78
Presbyterian Hospital, New York . . . . .	112	48
University College Hospital, London . . . . .	123	37
Total . . . . .	499	341

From these figures hemorrhage appears to be considerably more frequent, but as Gowers points out this is probably due to the fact that as hemorrhage is more often quickly fatal than softening, more such cases would be expected to die in hospitals. He thinks it probable that softening is in general the more common lesion. Clinically, the probable diagnosis of cerebral softening, in the writer's experience, has been much more often justified than that of hemorrhage.

The *character* of the apoplectic stroke itself gives some indication as to its cause. Premonitory symptoms when they have been present for two or three days, especially if there is a history of several slight previous cerebral attacks, suggest the gradual onset of thrombosis. In cerebral hemorrhage slight symptoms not infrequently precede the culmination of the attack, but do so for a much shorter period, from a few minutes to a few hours, and there may be the history of a previous apoplexy of considerable severity, but not commonly of repeated slight attacks. Hemorrhage may occur, however, after several thrombotic attacks.

The *occupation* of the patient at the time of the stroke may give some indication, for hemorrhage is to be expected when the circulation is active and thrombosis under the opposite conditions, when the circulation is depressed. Although it is uncommon to find a history of severe physical effort at the time of a cerebral hemorrhage, still more cases occur during the active hours of the day, and rest in bed and sleep seem to protect to a certain extent against this occurrence. Thrombosis is more common during sleep and rest in bed than hemorrhage, and this probably accounts for the preponderance of cerebral softening found at autopsy in hospitals where old chronic cases are treated. If one could determine exactly the state of the circulation at the time of the apoplexy it would be a great help in the diagnosis between these two lesions.

The essential thing to know is the circulatory condition at the site of the lesion, but the condition of the peripheral circulation is an imperfect index of this, especially if nothing is known of the patient previous to the attack. High blood-pressure is a frequent accompaniment of advanced arteriosclerosis, and may precede and accompany thrombosis as well as hemorrhage and should not be given too much weight. On the other hand, a feeble, soft pulse, with a weak, irregular heart, points strongly to thrombosis.

*Coma* is much more frequently absent in thrombosis than in hemorrhage, and when it develops rapidly it strongly suggests the occurrence of the rupture of a vessel. This is particularly so when the focal symptoms are not pronounced. On the other hand, a widespread paralysis, which has developed with slight or transient disturbances of consciousness, is usually due to thrombosis. However, there are many cases of thrombosis which have the typical development of hemorrhage, as well as a group of hemorrhage cases that have the onset typical of thrombosis.

A marked fall in the bodily temperature immediately after the apoplectic stroke speaks for hemorrhage, as do a rapidly rising blood-pressure, Cheyne-Stokes respiration, and the other signs of increased intracranial pressure.

*Convulsions*, particularly if repeated, point to an irritative lesion in or near the cortex, and are much more frequently associated with softening than with hemorrhage, and more often with embolism than with thrombosis. Cerebral softening bears the same relation to localized paralyses—monoplegias, the different types of aphasia, hemianopia, etc. Hemiplegia is the usual result of hemorrhage, but is also common with softening.

*Hemorrhage*, in the great majority of instances, is from some one of the central nutrient arteries, and involves the basal ganglia and internal capsule. This was true in 19 of the 27 lesions analyzed by the writer. The other lesions were situated as follows: twice in the frontal, twice in the occipital, and once in the temporal lobe. The cerebellum was involved twice, once with the pons, and in one instance the clot was in the pons and medulla. In Ellis' 31 cases it was in the region of the basal ganglia 26 times, not designated once, and was once each in the following situation: occipital lobe, parietal lobe and postcentral gyrus, cerebellum and pons. Hemorrhages into the pons are more frequent than those in the cerebellum (v. Monakow). True cortical hemorrhages are rare and are usually small.

*Softening* frequently involves the cerebral cortex; 20 of my 30 cases were so localized, and the basal ganglia and internal capsule showed softening 8 times, and in 2 instances the focus was in the lateral aspect of the medulla. Ludlum's experience was different. He found among 69 areas of softening only 9 in the cortex, all the others being in the neighborhood of the internal capsule. Thrombosis may occur anywhere, but is more frequent in the distribution of the middle cerebral artery and in that of the posterior cerebral. Emboli, in the great majority of cases, are lodged in the middle cerebral artery and its branches.

Therefore it appears that when the symptoms indicate a capsular lesion the probability is somewhat, although not much, greater that it is hemorrhage, but when they indicate a lesion in some other situation, the cortex particularly, it is apt to be softening. The examinations of the blood serum and spinal fluid may help to distinguish hemorrhage from softening. Thus a positive Wassermann reaction and the other signs of syphilis speak for thrombosis and softening. Blood or a yellow or reddish-brown color in the spinal fluid indicates hemorrhage.<sup>1</sup> P.

<sup>1</sup> Bigelow, *Cleveland Med. Jour.*, 1913, xii, 265.

Marie and Léri<sup>1</sup> noted that in cases of hemorrhage the blood serum has a fluorescent, greenish appearance not seen in softening. This appears shortly after the stroke and may persist for a week.

**Prognosis.**—The diagnosis of a cerebral vascular lesion always entails a serious prognosis, not necessarily as to the outcome of the present attack, but very generally as to the underlying vascular condition, which must remain a source of constant danger. The chief exceptions are those accidental cases occurring in the young in relation to acute infectious diseases, parturition, etc., and of certain cases due to syphilitic endarteritis which is amenable to treatment.

The great majority of apoplectic attacks are recovered from. In my own list there are the histories of 740 patients, most of whom had recovered from the attack and applied for relief of the residual paralysis. There were 448 of these. The fatal cases in the present list were among those treated in the hospital, and number 97 out of a total of 292 hospital cases. This includes a number in which the apoplexy was a contributory and not a direct cause of death. These figures give no idea of the relative frequency of death. The dispensary cases were all, of course, drawn from those that recovered, and there is no means of ascertaining the fatal cases of apoplexy occurring in the same community. The percentage of deaths (33.2 per cent.) among the hospital patients must be much greater than that which obtains in general, for it applies to a special group composed largely of the more severe cases or those complicated by other conditions.

In many cases previous attacks had occurred; usually one previous attack, not infrequently two and in several instances there were histories of three or more definite strokes. The time between the attacks may be short. It is usually estimated in months, and often there is an interval of years. The longest interval was sixteen years between the first and last attack. In this case the second attack occurred in two years, but there was an interval of thirteen years between the second and third and two between that and the fourth. The idea that the third attack is fatal has often been refuted. Among our cases, 79 died in the first, 10 in the second, 4 in the third, 1 in the fourth, and 3 after several attacks.

The gravity of the prognosis of any attack increases with the suddenness with which the coma has occurred, with its depth and with its duration. If the coma shows no sign of lessening after twenty-four hours the prognosis is very grave and becomes more and more grave the longer the coma persists. Patients have been known to recover from coma which has lasted for a long time, even up to six weeks. The writer saw a woman, nearly eighty years old, who had been in coma for three days, return suddenly to perfect consciousness as from sleep, much to the surprise of her watching relatives and to the chagrin of her physicians, who had given an absolutely hopeless prognosis. If the coma, at first slight, becomes later more pronounced, or returns after the patient has regained consciousness, it is of more serious import that the same degree of unconsciousness at or shortly after the onset of the attack.

<sup>1</sup> *Bull. de l'Académie de Méd.*, 1914, lxxi, No. 22.

When the initial fall of temperature, common in cerebral hemorrhage, persists and increases, it is of very grave import. On the other hand, when the temperature rises quickly, or the fever lasts longer than the first two or three days, the outlook is grave, and if at any time the temperature reaches great heights, it is almost always a signal of approaching death.

Marked changes in the respiration, particularly when it assumes the Cheyne-Stokes type, are of grave importance as indicating embarrassment of the respiratory centre. Cheyne-Stokes respiration, however, does not always mean immediate death; it may persist or come and go during many days. It may also have been long present before the apoplexy in association with cardiac or renal disease.

Rapidly rising blood-pressure as indicating increasing intracranial pressure is, of course, a serious symptom, but a constantly falling blood-pressure from a previously high level is one of the surest signs of approaching death. Cardiac failure has its usual significance. Any symptoms pointing to involvement of the lungs are very important, for many cases of apoplexy die of a terminal pneumonia.

An increasing swelling of the optic nerves is a serious symptom as indicating an increasing intracranial pressure, but it may be in association with uremic cedema of the brain and may entirely subside.

Any estimate as to the duration of an attack is very uncertain. The most rapidly fatal cases usually live for several hours, and a patient may survive a large hemorrhage for several days, even when it has ruptured into the ventricle. The time of death varies, among other things, with the character of the lesion, and Jones<sup>1</sup> gives an interesting table compiled from cases proved by autopsy:

Lesion.	Number of cases collected.	Percentage dying within twenty-four hours.	Within a week.	Within a month.
Hemorrhage . . . .	828	30.4	63.8	79.4
Thrombosis . . . .	158	15.8	38.0	74.7
Embolism . . . .	273	8.0	35.5	56.0

In our experience the average time of death in all fatal cases when this could be determined was about eight days. For the cases proved by autopsy it was 6.2 days; for cerebral hemorrhage, 5.3 days, and for cerebral softening, 7.5 days. The few cases in which the thrombosis was secondary to embolism had a somewhat longer duration—nine days.

The age and general state of health have also an influence on the prognosis. Young, strong individuals often survive a severe cerebral shock that would be rapidly fatal at a later period of life. On the other hand, in elderly people whose arteries are badly diseased, usually with associated cardiac and kidney trouble, a very slight stroke may be the beginning of a spreading thrombosis, and the patients become duller, lapse into coma, and die after a protracted illness.

**Treatment.—Prophylactic.**—This should begin with the prevention of the conditions liable to bring about vascular degenerations. As our knowledge increases as to the cause of arteriosclerosis, it is probable that

<sup>1</sup> *Brain*, 1905, xxviii, 546.

we will know better how to ward off or delay its occurrence. The enormous part that syphilis plays in the etiology of the apoplexies of early adult life makes the importance of its prevention and treatment too obvious for comment.

When vascular conditions are present that make the rupture or occlusion of an artery probable, we can, unfortunately, do but little to restore the integrity of the vessel wall, except when the change is syphilitic. When there is any evidence of involvement of the cerebral arteries in individuals under forty, syphilis should be suspected and thoroughly treated, even when there is no history and its possibility seems unlikely.

Patients with damaged bloodvessels should avoid excesses of all kinds, not only those of alcohol and tobacco, etc., but also excessive mental or physical labor. The diet should be light, the bowels kept freely open, and the other measures recommended for arteriosclerosis should be employed. If such measures do not cause a reduction in the blood-pressure when it is high, it is doubtful whether vasodilators are of any permanent value. Apoplexies occur during every conceivable activity of daily life, and so it is inadvisable to restrict the patient within too narrow limits.

**Apoplexy.**—Here as elsewhere effective treatment must depend upon an accurate diagnosis. In this instance similar symptoms may be due to vascular lesions of exactly opposite character—on the one hand the rupture of an artery, and on the other its occlusion, conditions which indicate quite different procedures if they are to be met directly. It is impossible in many cases to be certain which of these lesions is present, and the treatment must be such as would be applicable in either condition. It is often of the utmost importance to decide whether active measures should be employed in the endeavor to influence directly the initial lesion, especially whether the patient should be bled or undergo a surgical operation, or whether the treatment should be conservative.

**Bleeding.**—If the patient be seen immediately after the stroke, and the symptoms all indicate the bursting of an artery (a rapidly developing coma, lowered bodily temperature, a strongly acting heart, with a full, bounding pulse), and we have reason to believe that the high blood-pressure preceded and did not follow the hemorrhage, bleeding from a peripheral vessel might be indicated. This would be done in the endeavor to so lower the pressure inside of the ruptured vessel that the hemorrhage would stop and the extravasated blood be allowed to clot and further destruction of the nervous tissue be prevented. It must be remembered that the symptoms of apoplexy are not all dependent upon the local effect of the extravasated blood, and that many of them, coma itself among the rest, are often believed to be due to reflex effects, among which anemia of the cortex is of great importance. If this be true, lowering of the arterial pressure could hardly be expected to help.

High blood-pressure is by no means of itself an indication for blood-letting, for this may be a result and not a cause of the hemorrhage, as when the blood-pressure continues to rise, and there are other evidences of undue pressure on the medulla (Cheyne-Stokes respiration, etc.). Under these circumstances bleeding would seem to be absolutely contra-

indicated. If the apoplexy be due to the occlusion of a large bloodvessel, and this be associated with high blood-pressure, there can hardly be an excuse for venesection.

When we remember how rarely it is possible to be certain that we are dealing with a ruptured and not with an occluded vessel, and even if we be sure that a hemorrhage has occurred, how uncertain the indications for the rapid lowering of the general blood-pressure are, and, indeed, how incomplete our knowledge is of the effects of bleeding on the cerebral condition, we will see how seldom we have a definite indication for bloodletting in apoplexy.

All the recent authors refer to bleeding, but there is very little agreement as to the benefit which follows it, and it is difficult to find exact observations. von Monakow finds little reason, theoretically or practically, for its employment, and Cushing speaks strongly against its use, but Gowers, Starr, Oppenheim, and others recommend it more or less strongly. Goldscheider,<sup>1</sup> after considering the theoretical objections, concludes that these have been pushed too far, that they do not stand on a firm basis of known facts, and that although it must be acknowledged that permanent good effects are extremely rarely seen after bleeding in severe apoplexies, still he believes that it is indicated when the diagnosis of hemorrhage is certain and when the face is congested and the pulse is full and hard. He would bleed not only at the onset of the attack, but even later if coma had persisted with symptoms of hyperemia, or if the symptoms, which were at first slight, became more pronounced. In our own cases in which it has been used the writer has not been able to see evidence of the least permanent benefit. Even the blood-pressure, although it is reduced during the operation, rises rapidly again to near its original height. Only the most urgent cases were bled, however, and this may account for the lack of good results.

**Surgical Interference.**—Spencer and Horsley suggested, in 1889, that hemorrhage from the middle cerebral artery might be controlled by compression of the carotid artery, this being applied by digital pressure, either through the skin or upon the exposed vessel, or by permanent ligature of the vessel. This apparently has never been carried out.

Cushing,<sup>2</sup> in 1903, urged that the clot be dealt with directly, and gave details of the surgical procedure. He bases this on the theory that the hemorrhage occurs rapidly, has ceased when the physician sees the patient, and that the immediate symptoms are, for the most part, due to increased intracranial pressure, which can best be relieved by opening the cranial cavity and removal of the clot. Cushing has operated on a number of desperate cases of capsular hemorrhage, and in several instances certain of the alarming symptoms improved markedly, at least for a time. The most certain effect was perhaps on excessively high blood-pressure when present. At times the change in blood-pressure is not so marked, and, indeed, a decompression may have little or no effect. This is particularly so when the blood-pressure has accompanied arteriosclerosis and preceded and not followed the apoplexy. That the clot itself

<sup>1</sup> *Deutsch. med. Wchnschr.*, 1907, xlviii, 1977.

<sup>2</sup> *Am. Jour. Med. Sc.*, 1903, cxxv, 1017; Keen's *System of Surgery*, iii, 215.

can be successfully evacuated without subjecting the brain to more trauma than is justified, awaits demonstration, although isolated cases have been reported that seem to suggest this possibility. A decompression is clearly indicated when the patient's life is endangered by rapidly increasing tension. Marie and his co-workers<sup>1</sup> called particular attention to this, and recommend that the decompression be done over the sound hemisphere, for they believe that coma occurs only when the sound hemisphere is compressed, and that it is much less dangerous to operate on this side than on the other. They consider as favorable cases for the procedure those in which the coma and other signs of increasing intracranial pressure develop after twenty-four to forty-eight hours.

A dangerous increase in intracranial pressure may also follow thrombosis and embolism, and a decompression is equally indicated in these cases. Cushing has operated on a number of such cases, in several of which the procedure seems to have saved the patients' lives. Cushing's<sup>2</sup> brilliant example in operating on certain intracranial hemorrhages of the newborn has been followed successfully by a number of surgeons, particularly in France and Germany. K. Henschel<sup>3</sup> reviews the subject and recommends as a combined diagnostic and therapeutic procedure, lumbar puncture with puncture of the anterior fontanel. If a subdural clot is found and can not be aspirated, the major Cushing operation is indicated. He refers to the reports of 16 cases operated on by Cushing's method, of which 7 were cured.

The physician usually sees the apoplectic patient after the damage has occurred, and far too often hears the history of procedures that could only have done harm. The belief is very general among the laity that the early symptoms of a cerebral lesion, drowsiness, tingling or weakness in the hand or leg, etc., can best be combated by physical exercise, and that to give way to them is to court disaster. Even physicians do not seem always to appreciate the importance of absolute rest to a patient who shows the evidences of a beginning hemorrhage or other vascular lesion.

If the patient be conscious he should be cautioned to make as little voluntary effort as possible, not even to speak, this especially if there are evidences of aphasia. No endeavor should be made to arouse an unconscious patient, and the physician should be careful to make his examination with as little disturbance to the patient as possible. The patient must not be moved more than is absolutely necessary, and it may be wise not even to transfer him to another room. Absolute tranquillity of the patient is insisted upon; he should be in a room protected from noises and sources of irritation. Only those necessary for the care of the patient should be permitted to enter. He should be in bed with the head slightly raised, special care being taken that the neck is not bent and that nothing interferes with the return of blood from the head. When the patient is profoundly unconscious and the breathing is stertorous, and, indeed, perhaps in general, he had better be placed on his

<sup>1</sup> *Bull. de l'Acad. de Méd.*, 1913, No. 36; *Presse méd.*, June 6, 1914, p. 429.

<sup>2</sup> *Amer. Jour. Med. Sc.*, 1905, cxxx, 563.

<sup>3</sup> *Zentralbl. f. Gynäk.*, xxxvii, 9, 925.

side, so that the tongue does not fall back and impede respiration. Bowles insists that the apoplectic patient should always be kept in the lateral position, the paralyzed side being down.

The state of the circulation is of utmost importance. If the heart be feeble and weak, it must be regulated. Stimulants, such as nitrous ether, camphor, caffeine, and small doses of digitalis, are recommended, and even alcohol in habitual drinkers. The opposite conditions, a strongly acting heart, high blood-pressure, etc., which may indicate bleeding or surgical interference, have been considered.

When a hemorrhage is reasonably certain, active purgation is almost universally employed, and even in thrombosis it is the custom, although here milder purgation is recommended. Goldscheider speaks strongly against this practice, for he doubts whether the good derived from the slight lowering of the blood-pressure, even if this occurs, counterbalances the danger from the disturbances incident to the procedure, and during the first day or two he would simply endeavor to relieve the lower bowel by the use of enemata, care being taken not to disturb the patient more than is necessary. There is often incontinence of urine but there may be retention, and the condition of the bladder must be carefully watched. If retention occurs the bladder should be emptied two or three times a day by a catheter.

Ice to the head has probably no influence on the circulation within the skull, and so the common habit of applying an ice cap is harmless, and as it appears to be grateful in certain cases it may be used. In hemorrhage, agents that increase the coagulability of the blood are indicated and in thrombosis those which have the opposite effect may be used (Gowers).

When the patient is in convulsions or very restless he must be kept quiet by the use of bromide, chloral, veronal, and other hypnotic drugs. Morphine, on account of its depressing effect on the respiratory centres, should be given only when absolutely necessary.

In all cases when there is a possibility that the lesion is thrombosis due to syphilitic disease of the bloodvessels, active measures must be employed—salvarsan intravenously, mercury by injections, and later mercurial inunctions, and iodide of potassium in increasing doses.

Directly after an apoplectic stroke the patient should not be disturbed by attempts at feeding. The lips may be moistened with water and the mouth cleared with a swab of gauze, saturated with some cleansing mouth wash. If the patient remains unconscious for a long time and appears to be becoming weak for lack of nourishment, nutrient enemata may be given. If these are not retained, infusions of salt solution may be given. When the patient is able to swallow (and this ability should be carefully determined by the physician) liquids in small quantities may be given after the first few hours. Even in the mildest cases the diet must be very light for the first few days, and then increased very gradually.

In slight attacks it is always difficult to keep the patient quiet, but even in the mildest cases the physician must insist upon a rest in bed for at least a week, and it would be safer for two.

**Sequels.**—Destruction of any part of the brain entails some loss of function, and when the focus is large, or so situated as to interfere with



an easily recognized function, we have symptoms. Such focal symptoms commonly follow vascular lesions within the brain, and not infrequently make up most of the clinical picture, and instead of apoplexy we have palsy, as the older writers would say. The symptoms vary with the position of the focus, and a knowledge of cerebral localization is necessary for a clear understanding of this relationship. Local symptoms follow hemorrhage more frequently than softening within the brain. This is due to the fact that hemorrhage usually occurs in the region of the internal capsule where it causes destruction of important tracts, whereas softening may be anywhere, of small extent involving a silent area.

In most cases of apoplexy from all causes, however, focal symptoms are present. Such may be the first sign or evident only when consciousness begins to return. These symptoms may be so transient that they can hardly depend upon a destruction of tissue, but must represent a loss of function due to some passing state—anemia or edema.

After an attack of apoplexy the patient may show little or no *intellectual weakness*, but usually a careful inquiry will reveal a distinct lowering of mental vigor. The patient is not the man he was before the stroke, but tires more easily, is petulant, emotional, and often depressed. He is often unable to control the expression of his emotions, and weeps tumultuously upon the slightest cause, or even when he may not be conscious of any sad feeling. Less commonly his laughter may be uncontrolled. Similar symptoms due to arteriosclerosis are often present in a less degree before the stroke, which appears to have merely intensified them and to have reduced the patient to a lower intellectual level. When the lesion is a large one, or associated with widespread arterial disease, the defect may amount to almost complete dementia. On the other hand, especially in young subjects, a severe stroke may be survived with little effect on the mental functions. Pasteur, who for days was thought to be dying from an apoplexy when forty-six years old, subsequently did much of his brilliant work, and lived to be seventy-three.

If the lesion be so situated that it involves the speech area, *aphasia* follows and may be permanent. This is often associated with a marked lowering of the intellect, but at times this is not evident.

The physical defect that most often remains is *hemiplegia*, which results whenever the pyramidal tract is implicated, as in the common lesion of the internal capsule, or in any other part of its course. In severe cases the paralysis is widespread, affecting generally all of the movements of the opposite side of the body. The arm and leg are motionless, and often the facial muscles are powerless, and the common statement that one side of the body is completely paralyzed seem justified. A closer examination shows that this statement is far too general, and that there are many movements on the affected side which are little, if at all, implicated, and that those affected are so to an unequal extent. This attracted the attention of the older observers, and various hypotheses were advanced to explain it. The most widely accepted one was that which Broadbent<sup>1</sup> advanced in 1866, when he pointed out that the

<sup>1</sup> *Brit. and For. Med.-Chir. Rev.*, reprinted in *Selections from the Writings, Medical and Neurological, of Sir Wm. Broadbent*, London, 1908.

muscles which escaped were those which acted more or less constantly in association with corresponding muscles of the opposite side of the body, *i. e.*, bilaterally acting muscles, and that the degree of paralysis, when present, corresponded to the degree of their unilateral activity. This he explained by conceiving a more or less intimate association between the lower spinal nuclei. When muscles acted always together the connection was so close as to practically make one nucleus, which could be stimulated equally well from the right or left cerebrum, *i. e.*, its movements, composed of both right- and left-sided muscles, were represented on both sides of the brain, and a lesion on one side of the brain would not abolish them. When muscles, although usually acting together, could act unilaterally, the connection between the lower nuclei was less intimate, and their bilateral cerebral representation less complete, so that a unilateral lesion would paralyze them more or less, depending upon the degree of their unilateral activity. As constant bilaterally acting muscles, Broadbent considered those of the eye, back, neck, and chest. He regarded the upper facial muscles as being largely, but not completely, bilaterally acting muscles; the lower facial muscles he thought were almost entirely unilaterally acting. The muscles of mastication and of the tongue he also classed among those which act both symmetrically and unilaterally. Subsequent investigations have confirmed Broadbent's observations and his explanation is generally accepted as fundamentally true.

That movements and not individual muscles are represented in the brain (Hughlings Jackson) and that it is these which are affected in hemiplegia, is nowhere better seen than in relation to the external muscles of the eye. These muscles are never individually paralyzed, but the conjugate movements to the right or to the left are not infrequently affected, particularly in the early stage of a hemiplegia. In each of these movements muscles on the two sides of the body are involved. The rapid recovery of these movements cannot easily be explained on Broadbent's hypothesis, for although using muscles on the two sides of the body, the movements are themselves unilateral. It may be that these movements are represented in more than one part of the cortex, anteriorly in the motor cortex and posteriorly in the occipital lobe, and therefore one lesion would not be likely to destroy the pyramidal tract and the path from the occipital lobe.

The condition of the muscles of the eye in hemiplegia has been carefully reviewed. Mirallie and Desclaux tested the strength of the muscles by their power to overcome prisms, and came to the conclusion that in every case of organic hemiplegia, in which the face was the least involved, there was a weakness of all the muscles of the eye on that side, and also to a less extent on the sound side. S. A. K. Wilson,<sup>1</sup> working with Marie's material at the Bicêtre, and employing the same methods, came to exactly the opposite conclusion, *i. e.*, he was unable to determine any weakness of the external muscles of the eye in cases of hemiplegia beyond that which was often found in healthy individuals.

<sup>1</sup> *Rev. Neurol. and Psychiat.*, 1904, p. 265.

*Ptosis* due to weakness of the levator palpebræ superior is at times observed with lesions of the cerebral hemisphere, and the involvement of this muscle in hemiplegia is cited as an exception to the general exemption of the muscles supplied by the third nerve. Ptosis is, however, rare from a lesion within the cerebral hemisphere, and Wilbrand and Saenger, in their careful analysis of the literature, were able to find the records of very few satisfactory cases. When the lesion is in the brain stem and can directly implicate the nuclei of the ocular muscles, their paralysis is, of course, a common symptom.

The movements of *mastication* on the paralyzed side are more or less weak. Mirallie and Gendron found that the weakness was particularly of the masseters and pterygoids, and more marked the more recent the case of hemiplegia.

Although Broadbent recognized that all the facial muscles were implicated in varying degrees in the paralysis of hemiplegia, those about the eye being less affected, later observers applied his hypothesis more strictly, and the statement that the upper facial muscles were completely spared in a lesion of the brain, received general acceptance. A review of the question by a number of writers has confirmed Broadbent's original statement in a remarkable way. In early cases, when the face is at all implicated, it is usually so in its entirety. The movements of the upper facial muscles, in a great majority of cases, recover rapidly, and after a few days their weakness is revealed only by careful examination. It may be noticed that the eyebrow on the paralyzed side is a little lower than on the other, that the movements of the forehead, although present, are weaker than those on the other side, and that the eye cannot be closed so quickly and firmly. The patient is unable to close voluntarily the eye on the paralyzed side without at the same time closing the opposite eye, even when he may have been able to do so before the stroke, and this may be the only remaining evidence of the weakness of this group of muscles (the orbicularis sign). The paralysis of the muscles about the mouth (the lower facial group) is much more permanent, and in some cases a slight drooping of the corner of the mouth during voluntary effort, may be the last evidence of a hemiplegia. Although this relation between the degree of paralysis of the upper and lower facial muscles holds good for the majority of cases, exceptional cases have been reported in which the condition is the reverse, the upper group being more paralyzed than the lower.

The movements of the *larynx* are as typically bilateral as any in the body. Horsley and Beevor found that stimulation of the cortex or the internal capsule of either hemisphere caused movements of both vocal cords. In hemiplegia these movements are not permanently affected, although some involvement of them probably takes place in the difficulty in speech so common in the first few days after an apoplexy.

The movements of the *soft palate* are rarely referred to in detail in relation to hemiplegias, but Tetzner<sup>1</sup> studied this question and finds that the condition varies greatly. In certain cases there is complete paralysis,

<sup>1</sup> *Neurol. Centralbl.*, 1909, p. 520.

and in others the palate moves only during gagging and then usually symmetrically. In a number of cases, fifty-two, the palate moved during phonation, and in those he found the movements symmetrical, the soft palate in front and above the anterior arch being drawn backward and toward the paralyzed side, making the picture of a tent, whose point was directed toward the paralyzed side. In a few cases this movement of the palate was the only remaining sign of the paralysis.

The *tongue* when protruded not infrequently deviates toward the paralyzed side. This is usually explained by the statement that the muscles of that side, being weak, are overbalanced by their opponents, and when the tongue is protruded it is pushed over toward the weak side. Beevor<sup>1</sup> showed that the condition is not so simple, and pointed out that stimulation of the cortex reveals two separate centres for the movements of the tongue, one in the upper part of the tongue area, which causes deflection of the tongue toward the opposite side, and another situated lower and more anterior, the stimulation of which causes protrusion of the tongue in the mid-line. This latter affects equally the movements on the two sides of the tongue. It is some disturbance of this movement, protrusion of the tongue in the median line, that causes the deflection of the tongue toward the hemiplegic side, and not the voluntary movement of turning the tongue toward that side. This latter is a unilateral movement, and Beevor showed that it is affected in hemiplegia, the patient having more difficulty in putting the tongue into the cheek on the paralyzed side than in moving it in the opposite direction.

The explanation of the deflection of the tongue when the patient endeavors to protrude it straight is not clear, but Beevor advanced the tentative theory that although each hemisphere contains the movements for both sides, still for a perfectly balanced movement the action of both hemispheres is required, and a lesion in one causes a greater amount of weakness in the movements of the opposite side. This amounts to an elaboration of the common statement.<sup>2</sup>

The movements of the mouth, pharynx, larynx, tongue, etc., are used in many most important complex functions, deglutition and speech, for instance. A single lesion above the medulla does not often permanently disturb the power of swallowing, nor, unless in the speech sphere, abolish the movements underlying speech, although in rare instances such permanent effects occur and result in a typical pseudo-bulbar paralysis. Panski<sup>3</sup> refers to such cases as well as to some other unusual symptoms in hemiplegia. Pseudo-bulbar paralysis usually follows two apoplectic strokes, first on one side and then on the other, implicating the fibres from both cortices to the bulbar nuclei. The lesions are most often in the internal capsule.

The movements of *respiration* are bilateral, and so little affected in hemiplegia that in the records of the routine examination they are very

<sup>1</sup> *Brain*, 1906, xxix, 487.

<sup>2</sup> The movements of the tongue following stimulation of the twelfth nucleus, root and nerve, are the subject of a study by Mussen, *Brain*, 1909, xxxii, 203.

<sup>3</sup> *Deutsch. Zeitschr. f. Nervenh.*, 1914, li, 1.

generally noted as being equal. In a number of cases where special attention has been called to this point no difference in expansion of the two sides of the chest could be determined. At times, however, there is a marked difference, the movements on the paralyzed side being less than those of the opposite side. Hughlings Jackson<sup>1</sup> thought that the condition differed in quiet and forced breathing; during quiet breathing the movements on the paralyzed side being greater, while in voluntary or forced breathing they were less than those on the opposite side. This was confirmed by a number of investigators, but West<sup>2</sup> found no such difference. In his carefully studied case the movements were less on the paralyzed side during every character of respiration, and the chest on that side actually bulged during the strong effort of coughing.

The muscles of the trunk act both bilaterally, as in forward and backward flexion of the body, and also unilaterally, in the movements toward the right and left. These movements are affected differently in hemiplegia, those acting bilaterally being but little affected, the muscles on the paralyzed side acting nearly as strongly, although somewhat later than their fellows. The lateral movements may be quite differently affected. Beevor<sup>3</sup> reported a case of left hemiplegia which again shows that movements and not muscles are represented in the cerebrum. When sitting, the patient could not lean to the left with as much force as to the right, although he showed a tendency to fall toward the left, *i. e.*, there was a weakness both in the movements of the muscles which bend the body to the left (left-sided trunk muscles), and also those which keep it from falling in that direction (right-sided trunk muscles); both these movements Beevor considered as left-sided movements. These same muscles acted strongly in right-sided movements, *i. e.*, the patient could bend himself strongly to the right (right trunk muscles) and could from that position bring himself with normal power to the upright position (left trunk muscles).

All movements of both limbs are abolished at first in a severe hemiplegia, but as the patient begins to recover, or from the first, in slight cases, the tendency for the arm to be affected more than the leg is apparent, and movements return first in the lower limbs. If the lesion be sharply localized well back in the internal capsule or involve the leg centres in the cortex, the paralysis is, of course, most marked in the leg; but the above statement is true for the great majority of hemiplegias, and it is a very uncommon case in which there is no return of motion in the leg, and generally there is some return of movement in the arm.

As the movements begin to return there is evidence of a certain selection. This has been the subject of a number of investigations, and here again we find that it is rarely individual muscles that are paralyzed in hemiplegia, but movements which require the action of many muscles. Of these muscular mechanisms there are some which are more apt to remain paralyzed and others which show a tendency to recover. In the leg the groups that are most affected are, in a general way, those which advance the leg during walking, *i. e.*, the flexors, abductors, and external

<sup>1</sup> *Lancet*, 1905, i, 476.

<sup>2</sup> *Quart. Jour. Med.*, 1907-1908, i, 448.

<sup>3</sup> *Brit. Med. Jour.*, 1909, 881.

rotators of the hip and the flexors of the knee and the dorsal flexors of the ankle; whereas the opposite group of muscles is little affected and recovers first. In the arm the condition seems to be more complicated. The movement which opposes the thumb to the little finger is most apt to remain paralyzed. Opening of the hand and outward rotation of the forearm are more affected than closing the hand and inward rotation. Extension of the elbow is more affected than flexion. The movements forward and backward of the upper arm and all the movements of the shoulder girdle are usually lost. The disability is not always proportionate to the return of muscular strength, and at times one finds an arm in which there is little or no decrease in the force of the individual movements, but which is practically useless for all its finer activities.

It is apparent that the varying degrees of paralysis following a cerebral lesion cannot be entirely explained by Broadbent's simple hypothesis of the close association of the lower centres for bilaterally acting muscles. The condition is much more complicated; the association may be between the cerebral centres themselves, as seems to be the case for the trunk muscles, which, although unilaterally represented in the cortex, as shown by simulation experiments, act symmetrically and are little paralyzed from a unilateral lesion. Förster<sup>1</sup> believes that the selective paralysis of the limbs is due to an innervation of the retained muscles by auxiliary fibres which arise from the homolateral motor cortex and run as direct pyramidal fibres both in the anterior and lateral tracts of the cord. Röthmann,<sup>2</sup> in a review of the subject finds no proof of the existence of such fibres, but believes that the retained movements are due to the renewed activities of phylogenetically old, sub-cortical, motor centres—chiefly the red nuclei and spinal centres—as modified by the upright position in man. A full explanation must wait for a more complete knowledge of the physiology of motion.

The movements on the non-paralyzed side are usually somewhat affected, *i. e.*, their force is reduced. Brown-Séquard called attention to this in 1882, and Sternberg<sup>3</sup> has made a study of the question. Often the reduction in the power is not great, but at times it is marked. The force with which a movement can be executed on the paralyzed side is generally increased, if at the same time the patient makes the corresponding movement on the opposite side. The effect of simultaneous movements on the normal limbs varies but it is never very great.

Allied to this reinforcement during simultaneous movements of the two sides are the *associated movements* which are often seen in cases of hemiplegia. When a strong effort is made with the non-paralyzed side it is very usual to see some involuntary movement of the corresponding paralyzed limb. During efforts to make a definite localized movement on the paralyzed side, as closing the hand, flexing the ankle, etc., the whole limb may be brought into play, and, indeed, the other limb on the same side may be moved. It is also not very infrequent to see associated symmetrical movements of the opposite non-paralyzed side. These phenomena are particularly well seen in infantile hemiplegia. Strümpell

<sup>1</sup> *Deutsch. Ztschr. f. Nervenhe.*, 1909, xxxvii, 349.

<sup>2</sup> *Ibid.*, 1914, l, 406.

<sup>3</sup> *Ibid.*, 1908, xxxiv, 128.

pointed out that if a hemiplegic, or, indeed, any patient in whom the pyramidal tract was affected, be placed in a sitting position and asked to flex the paralyzed hip while the observer resisted the effort by making downward pressure on the knee, there will be a strong involuntary action of the tibialis anticus muscle, even though this muscle may be paralyzed to voluntary effort (Strümpell's tibialis phenomenon). This is a good example of an associated movement, and illustrates the inability of a hemiplegic to make isolated movements. During certain involuntary or reflex movements the paralyzed limbs may be moved in a surprising manner; thus, during yawning the arm may be forcibly raised and drawn across the chest.

Many explanations have been advanced to account for these associated movements. They are obviously due to a spread of the impulse beyond the centres which control the intended movements. This is believed to be due either to a hyperexcitability of the lower centres or to an increase in the stimulus, *i. e.*, voluntary effort, the direction of the spread depending upon associated paths between these centres. These connections may be the remains of primary paths laid down in the early evolutionary history at a time when all movements were bilateral, and which have persisted to a greater or less extent during development (phylogenetically old mechanisms).

In newborn children the tendency to associated movements is very great, and becomes less as the facility to make coördinated movements is acquired. Later in life, when new, highly specialized movements are being learned, writing, dancing, etc., at first many more movements are brought into play than are required for the action. During practice the unnecessary movements are inhibited, the necessary ones are emphasized, and finally, when the movements are perfectly learned, only necessary muscles are moved, and only with the required amount of energy. The processes underlying this must be very intricate, but an essential factor is the cerebral control, acting, for the most part, through the pyramidal tracts, and an interruption of one of these tracts, as in apoplexy, causes a loss of the finely coördinated movements, and a reversion to a more fundamental type. The importance of the sensory factor is obvious.

The *muscular tension* in the paralyzed limbs is, in the great majority of cases, at first very much reduced, but if the lesion is situated in or near the cortex, and is irritative, the limbs may at once become rigid, or be the seat of recurrent convulsions. This is the early rigidity to which Todd called special attention. These irritative phenomena usually pass off in a few days, but are at times said to persist and to pass over into the state of late rigidity with permanent contractures.

In practically every case of hemiplegia in which there is a certain degree of recovery a condition of muscular *rigidity* develops in the paralyzed limbs (late rigidity of Todd). This begins when there is some evidence of returning voluntary power, and at a time when the deep reflexes have already become exaggerated. The degree of contracture is not exactly proportionate to either of these factors. It is most marked in hemiplegias in which the return of voluntary power has been but slight. The limbs

become rigid and tend to assume definite positions. The arm is adducted, flexed at the elbow, the thumb and fingers being flexed into the palm. The leg is straightened, *i. e.*, the contracture is most marked in the extensors of the hip and knee and in the plantar flexors of the hip. The opposite condition, the arm in extension and the leg in flexion, is very exceptional. The muscles most contracted are, in a general way, those in which the recovery of voluntary power has been greatest, and this relative overstimulation of certain muscles has been given as an explanation of the contracture. The condition, however, is far more complicated, and various theories have been advanced to account for it. Here, as in associated movements, we have the action of a portion of the central nervous system from which an important controlling influence has been taken.

In the *contractures* of hemiplegia there is a condition of increased muscular tension, *i. e.*, of heightened muscular tonus. Muscular tone depends on many things, but peripheral sensory impressions are necessary, *i. e.*, it is essentially a reflex phenomenon. That the cerebral hemisphere has a modifying influence on these centres is well established, and the remarkable effect upon the muscular tension of the withdrawal of these influences is beautifully illustrated by Sherrington's "Decerebrate Rigidity." In animals whose cerebrum is cut away the muscles show a remarkable state of hypertension, being maintained in a flexed position against gravity. If, however, in such an animal the posterior roots which contain the sensory fibres from a leg be cut, the limb becomes perfectly flaccid. This seems quite analogous to the fact that contracture does not occur in a hemiplegic with tabetic involvement of the posterior roots and gives a physiological basis for Förster's procedure of cutting the posterior roots as a treatment of the hemiplegic contractures.

The positions that the limbs assume in the hemiplegic with late rigidity appear to depend, at least in part, upon the positions in which they are maintained, and can mechanically, by the application of splints, be altered. The attitude of the leg in bed, as determined by gravity, etc., is an important factor in its extensor contracture. When the origin and insertion of a muscle are, either actively or passively, brought closer together or separated, there is normally a reflex tendency to maintain the muscle in that position (fixation reflex). Sherrington studied this plastic tonus in animals during decerebrate rigidity and under other conditions, and gives evidence to show that it depends upon definite reflex phenomena. This tendency is much exaggerated when the cerebral control has been withdrawn, so that when a hemiplegic contracts a muscle there is a marked tendency for the contraction to be maintained, and this explains why the contracted muscles are, as a rule, those muscles in which voluntary power has to some extent returned. At first the contracture is entirely active and subsides during sleep and under an anesthetic, but after it has been maintained for a certain length of time anatomical changes occur in the muscles, so that they become permanently shortened and the contracture becomes fixed.

The *walk* of a hemiplegic patient is very characteristic, and is conditioned by the loss of voluntary power and the occurrence of contrac-



tures. The paralyzed leg is held rigidly in extension and usually rotated inward. As it is advanced the weight of the body is thrown over the sound limb and the paralyzed leg is swung forward in a curve, the toe often scraping the ground, the movement depending largely upon the muscles of the trunk. With every such movement there may be associated movements in the contracted arm.

*Exaggeration of the deep reflexes* is an early sign of the abnormal condition of irritability of the lower subcortical centres which follows interruption of the pyramidal fibres, and may occur soon after the apoplectic stroke. It is a constant accompaniment of the later hemiplegic state. On the paralyzed side the knee- and ankle-jerks are excessive, and clonus at both places can usually be obtained. An active abductor reflex of the thigh is common, and can be elicited, not only directly, but when the blow is struck on the opposite healthy leg, and it often accompanies the knee-jerk on that side—crossed adductor reflex. On the non-paralyzed side the deep reflexes are usually increased, but not to the same degree as those on the hemiplegic side. In the paralyzed arm there is an analogous exaggeration, the biceps and triceps reflexes are intense, the scapula reflex is marked, and at times a definite clonus at the wrist may be brought out. When contractures are extreme they may so mask the reflexes that it is difficult to elicit them.

The *skin reflexes* vary. Those from the conjunctiva and the abdominal wall usually remain absent. The reflexes from the skin of the leg and foot may be excessive, any irritation causing a widespread movement of the paralyzed leg. This is usually in flexion at the hip, knee, and ankle, and the marked dorsal flexion of the great toe appears to be a part of it. This latter movement, Babinski reflex and its modifications, is justly regarded as a most important evidence of a lesion in the pyramidal tract, and is very generally, although not constantly, present in cases of hemiplegia. Crocq,<sup>1</sup> who has for years studied the question of tonus, reflexes, and contractures, has given an interesting summary of his present view, and Walshe<sup>2</sup> investigated the reflexes clinically in the light of Sherrington's findings and showed how clearly they correspond.

Many voluntary movements of the paralyzed limbs are lost, but even those which are retained are, as a rule, performed in an awkward, inexact manner. At times, when there is but little loss of actual muscular strength, the inability to perform the finer movements is very marked, and there may be almost complete loss of function with but a slight decrease of muscular strength. Ataxia in its more limited sense occurs in those cases with marked and permanent loss of the muscle sense.

Movements of the affected limb may be accompanied by a definite *tremor*, which at times has the characteristics of that seen in paralysis agitans, and at times has more the volitional type, approaching the tremor of multiple sclerosis. Not infrequently, and more especially in cases early in life, the paralyzed limbs are in more or less constant involuntary motion. These movements are at times confined to the hand and foot, and consist in a slow, worm-like twisting and bending of the fingers,

<sup>1</sup> *L'Encephale*, 1914, ix, 147, 197, 293.

<sup>2</sup> *Brain*, 1914, xxxvii, 269.

associated with flexion and extension of the wrist, analogous movements occurring in the foot. Although in a typical case of *athetosis*, as this condition is called, the movements do not involve the whole limb, the muscles of which, however, are usually spastic, still not infrequently the rest of the limb may be affected by other spontaneous movements and may at times be jerked violently about.

Very similar movements may occur alone, and we then have a condition that has been called *posthemiplegic chorea*. Many authors make no distinction between these two conditions, and it is undoubtedly true that it is often difficult to decide in just which category a given case should be placed. Typical, uncomplicated, posthemiplegic chorea is much less frequently seen than athetosis, but when present it has marked, distinguishing characteristics. The movements are widespread, affecting the face, the body, and all the segments of the extremities. They are manifold, violent, and have a quick, trembling character. Contractures are not common. Both occur more frequently in infantile hemiplegias, and usually in those cases in which there has been a considerable return of voluntary power. Indeed, the limbs affected may show a little or no decrease in power. Various explanations have been given to account for the condition. It appears to occur most frequently when the lesion is in the neighborhood of the basilar nuclei, particularly of the optic thalamus or the subthalamic region, and the most probable explanation seems to be that it is some disturbance of the afferent paths, particularly those from the cerebellum, which run through the red nucleus,<sup>1</sup> and that lesions of the motor cortex and pyramidal tracts have but little to do with it.

In hemiplegia following apoplexy the nutrition of the muscles is, as a rule, not much affected, and never to the degree that is constantly present when the lesion is in the lower motor segment. When the attack has occurred in early youth there is generally a considerable retardation in the growth of the paralyzed side, and later in life the contrast between its size and that of the normal side is striking. This general wasting or retardation in the growth of a paralyzed limb is regarded by many as depending upon disuse, and is distinguished from the degenerative atrophy which depends upon a disturbance of the lower motor neurone. Others regard it as a trophic disturbance, and point in confirmation to the lack of development of the hair and breast which occasionally occurs on the paralyzed side.

A number of observers have pointed out that it is not very uncommon to see such a degree of muscular wasting occurring so quickly, following a cerebral lesion, as to suggest the degenerative type of muscular atrophy. Indeed, Steinert asserts that some degree of this type of atrophy occurs with every hemiplegia. It usually affects the muscles of the arm more than those of the leg, and reaches its maximum in a few weeks, after which it may recede, the wasted muscle regaining most of its lost volume. Associated with the atrophy he found a definite decrease in the electrical excitability of the nerves and muscles, some slowness of the response,

<sup>1</sup> Gordon Holmes, *Brain*, 1904, xxvii, 327.

and a tendency for the excitability to disappear after repeated stimulation (myasthenic reaction).

The *bladder*, the disturbance of which is so common during the apoplectic stroke, usually regains its function quickly, but in certain cases some permanent disability remains. The patient may have great difficulty in starting the flow or may be unable to restrain the bladder from emptying soon after the desire is felt.

*Loss of sensation* is much less common than motor paralysis as a permanent effect of a cerebral vascular lesion. In many cases the disturbance, if present, is so slight that it escapes notice, but it must be acknowledged that far too often the examination has been too superficial to discover the slighter grades of sensory loss. We have the record of 70 cases in the Johns Hopkins Hospital in which definite sensory disturbances were noted, and of 56 cases in which it was stated that there was no such loss. It is probable that in the majority of the other cases, where the records are silent in this regard, at least rough sensory tests were made and no disturbance found. Individual observers who have made special inquiry, have found some sensory loss in most cases of hemiplegia. Sandberg<sup>1</sup> examined 31 cases in Strümpell's clinic and in only 10 was he unable to discover any objective sensory disturbance, and among these were a number who complained that the impressions were not so acute from the paralyzed as from the normal side.

At times the apoplexy has been due to a focus so situated that the sensory loss is the chief, or the only symptom of a local destruction of the brain. Such cases are uncommon, and von Stauffenberg<sup>2</sup> was able to find in the literature only 8 cases of hemianesthesia without motor paralysis which were completed by autopsy. He included lesions of all kinds. The studies of Head and Holmes<sup>3</sup> have done much to clarify our knowledge of the subject. The sensory loss from cerebral lesions varies with the level of the sensory path involved. Below the optic thalamus the primary sense qualities of touch, pain, heat and cold, and those underlying postural and spacial discrimination, are subserved by the fibres of the fillet, and may all be disturbed by a lesion of this tract. In apoplexy such a lesion is usually near the thalamus, and very generally this structure is also involved, in which case there will result a marked lowering of sensation in some or all of its qualities, "deep" or muscular sense being constantly affected, combined with a peculiar, painful response, characteristic of thalamic lesions, *i. e.*, when any sensation is aroused it is accompanied by a most disagreeable, painful feeling, and, indeed, this may be the only response, the patient being quite unable to distinguish the character of the stimulus. The pain may be apparently spontaneous, occurring in paroxysms of great severity for many years. This is the distressing feature of the "syndrome thalamique" of Dejerine and Roussy.

In the optic thalamus there is a re-grouping of the sense qualities, and in it fibres arise to pass to various regions of the cerebral cortex. Lesions of the sensory cortex, post-central and parietal convolutions particu-

<sup>1</sup> *Deutsch. Ztschr. f. Nervenh.*, 1906, xxx, 149.

<sup>2</sup> *Arch. f. Psychiat.*, 1909, xlv, 683.

<sup>3</sup> *Brain*, 1911, xxxiv, 102.

larly, if large enough, cause sensory loss. The defects are largely of judgment, and do not usually involve fundamental sense qualities as touch, pain, and temperature, to any marked degree. The recognition of the position of a limb or its passive movements (muscular sense) is most frequently disturbed. Not infrequently a patient who is conscious of the most delicate touch is entirely unable to localize it, or to form an idea of the size, shape, weight or structure, of an object placed in his hand (astereognosis).

A lesion may involve the optic tract and the fibres which are leaving it, and then there will be combined sensory loss of the cortical type with the painful phenomena of a thalamic lesion.

A sharply localized lesion in the brain stem, as that following an occlusion of the posterior inferior cerebellar artery, frequently gives a dissociated sensory disturbance of the spinal type, absolute loss of pain and temperature, with perfect retention of touch and muscular sense.

The distribution of the *sensory* defect varies greatly. It may involve the whole side, but the mucous membranes and the skin of the face are usually spared, and even when most widespread the degree of loss differs, the distal portion of the limbs being more affected, and the hand being usually more involved than the leg. The anesthesia may be entirely confined to the distal portions of the limbs or of one limb or may be found in more or less isolated areas of the affected side. The division between normal and affected skin areas is not sharply marked, but one passes more or less gradually into the other. It has been shown that the areas of sensory loss, following a cerebral lesion, may have the distribution of those seen in lesions of the spinal cord.<sup>1</sup> This is particularly so when the lesion is in the cortex, and Russell and Horsley<sup>2</sup> suggest that this depends upon a re-representation of the spinal segments in the sensory cortex.

*Hemianopsia* of the opposite side occurs when the visual path is involved in a cerebral lesion, and is a frequent accompaniment of organic hemianesthesia. Total blindness of one eye due to optic atrophy, secondary to occlusion of the thalamic branch of the internal carotid artery, is a symptom of thrombosis of this vessel. The blind eye is on the side of the lesion, and not of the paralysis.

*Hearing* is not much impaired by a unilateral lesion. Impressions from each ear go to both sides of the brain, although probably in greater intensity to the opposite side. One would expect a certain degree of deafness in the opposite ear as compared with its fellow when the auditory centre or its afferent path is implicated. Such a loss has been frequently found from a lesion in the region of one of the posterior corpora quadrigemina.

*Vasomotor and Other Sympathetic Effects.*—During the apoplexy and directly afterward the paralyzed limbs are not infrequently warmer than those of the other side, and it is said they may be swollen and cedematous. Often the patients sweat profusely, but except in lesions of the brain stem there does not seem to be any difference between the two sides.

<sup>1</sup> Goldstein, *Neurol. Centralbl.*, 1909, p. 114.

<sup>2</sup> *Brain*, 1906, xxix, 137.

von Bechterew<sup>1</sup> and others have described unilateral abnormalities in the secretion of sweat following cerebral lesions, and have shown experimentally that definite effects in this secretion can be obtained by stimulating the cerebral cortex. In the chronic stage the paralyzed limbs are, as a rule, cooler than their fellows.

There appears to be no doubt that the cerebrum influences sympathetic activities, but little has been proved as to the manner and extent of this influence. The most striking case seen by the writer was in an infant sixteen months old, in whom there was a most remarkable pallor and coldness of the entire left side, which had been paralyzed during an acute cerebral attack. The vasoconstrictor symptoms and the paralysis cleared together in about six weeks.

In several cases, in which there was every reason to believe, and proved in two by autopsy, that the lesion was a softening of the lateral aspect of the medulla, following an occlusion of the posterior inferior cerebellar artery, there were marked evidences of paralysis of the sympathetic functions on the same side—slight ptosis, a pupil that dilated poorly in dim light, and an absence of sweating on that side of the body.

*Bedsore.*—In patients who are long in bed, especially when there is lack of control of the sphincters, bedsore tend to develop. Charcot believed this to be a definite trophic disturbance, but most observers regard it as the result of pressure, lack of cleanliness, and disturbed nutrition.

**Treatment of the Hemiplegia.**—The management of a patient after he has recovered from the cerebral attack must, in the first place, be directed against the occurrence of a new attack. These measures have been considered under the prophylactic treatment of apoplexy.

In the treatment of the hemiplegic state itself, the paralyzed limbs should be rubbed, moved passively, and the arm supported at the elbow in the endeavor to keep the shoulder from sagging, and so prevent the pain which is so often referred to this joint. The galvanic current applied directly through the joint sometimes is a benefit. Systematic massage, passive movement of the joint, and electrical stimulation of the muscles appear, at times, to be of use in reëstablishing motor power. When voluntary power begins to return this should be encouraged by practice, and the patient should have regular gymnastic exercises. When the tendency to contracture is very pronounced, one should endeavor to see that the limbs are stiffened in the most convenient positions, and fortunately this is the attitude that is most often assumed—the arm in flexion and the leg in extension. The usual methods employed are massage, passive movements, and at times the application of splints, but other methods have been suggested, and Criegern<sup>2</sup> exercises the arm after the blood has been driven from it by the use of the Esmarch bandage, the limb held above the head.

In cases in which the contractures in the limbs have become so intense as to prohibit their use, as not infrequently occurs in the diplegias of infants, Förster<sup>3</sup> devised, and Tietze executed, a most interesting surgical

<sup>1</sup> *Arch. f. Anat. u. Physiol.*, 1905, p. 297; *Neurol. Centrabl.*, 1907, p. 187.

<sup>2</sup> *Fortschr. d. Med.*, 1909, xxvii, 2.

<sup>3</sup> *Mitteil. aus d. Grenzgeb. d. Med. und Chir.*, 1909, xx, 493.

procedure. This is based on the theory that the spasticity is a reflex phenomenon, and depends largely on the afferent impressions which the lower centres receive from the limbs, and that if these could be abolished the limbs would become flaccid, and if they could be decreased in amount the spasticity would be lessened. Förster, considering the overlap in the distribution of the posterior roots, each area of the skin being supplied by at least three, and each muscle by two and usually three roots, concludes that two posterior roots supplying a definite muscle group might be safely cut if they were not neighboring roots, and that in this way the spasticity might be decreased with slight, if any, sensory loss. Stoffel, on the other hand, divides some of the fasciculi of the motor nerves going to contracted muscles, thus reducing their strength. The subsequent reëducational treatment is, however, a most essential part in both of these procedures, and must account in great measure for the results. Other more usual orthopedic measures of cutting and transplanting the tendons are sometimes useful.

## CHAPTER XIII.

### DISEASES OF THE CEREBRAL NERVES.

By E. W. TAYLOR, M.D.

**Introduction.**—The title, “Diseases of the Cerebral Nerves,” is somewhat misleading, first, because of the relatively brief statement necessitated by space, and secondly, because it is not possible to consider affections of the cranial nerves with any degree of completeness, irrespective of the general diseases of which they so often form a part. The attempt has been made in this section to consider only those disturbances which are characteristic of the individual nerves, and not to infringe on the domain of the ophthalmologist, otologist, laryngologist, or the broad field of the general physician or surgeon. Such inconsistencies as may occur are necessitated by the difficulty of classification. It is evident that many general diseases are in part manifested by disturbance on the part of various cranial nerves. An attempt has been made to allude to these frequent associations without entering into undue detail.

#### OLFACTORY (FIRST) NERVE AND TRACT.

**Etiology.**—Disturbances of smell may arise from lesions in any part of the course of the olfactory nerves or tracts from the peripheral distribution in the nose to the ultimate terminations in the brain. If, for example, the olfactory mucous membrane of the nose is affected through local irritation, swelling or any obstruction of the passage leading to this area, the sense of smell of necessity suffers, even though the nerve be not affected. Local disease may destroy the nerve endings in the olfactory area, naturally leading to a greater or less degree of anosmia. Actual injury of the olfactory nerves at the base of the brain, where for a short distance they lie in close relation to the bone, may produce a like disturbance. Such a lesion, though probably often overlooked, undoubtedly occurs as a result of contusion of the brain, or in extensive fractures of the base involving the frontal region. Loss of the sense of smell has been observed after an attack of influenza and in senile conditions. A *congenital* anosmia and a rare *hereditary* form have also been described. Various perversions of smell are not uncommon. A condition of increased sensitiveness, hyperosmia, has been described in hysteria, usually associated with altered appreciation of odors. This condition is, no doubt, analogous to the extreme acuity of vision occasionally met with in that disease. Subjective sensations of smell, parosmia, are, on the whole, rare, but undoubtedly occur in disease of the brain, or from irritation of the nerve in its extracerebral course. Olfactory hallucinations occur in certain forms of insanity, and may possibly be regarded as having some localizing significance. As an epileptic aura, also, such perversions of

smell may be significant. Gowers speaks of a case, reported by Urben, in which parosmia occurred in the course of tabes dorsalis.

**Pathology.**—The pathological anatomy of affections of the olfactory nerves is closely associated, on the one hand, with local disturbances in the nasal cavity, bearing merely a secondary relation to the nervous system, and, on the other hand, to traumatism of the cranial walls or the brain. Infections, notably meningitis, may include the olfactory apparatus in their spread, but usually in such cases other symptoms so far predominate that special disturbances of smell are overlooked. It should be remembered that a possible source of infection of the brain meninges may be through the cribriform plate of the ethmoid bone, particularly if it has sustained a fracture. Primary disease of the olfactory apparatus, apart from the conditions mentioned, is rare.

**Symptoms.**—The sense of smell, broadly considered, includes the recognition of odors through the anterior nares, stimulating the olfactory area, and the recognition of flavors through the posterior nares, also through the mediation of the olfactory nerve. Whether or not the fifth nerve contains elements which subserve the special sense of smell is open to doubt. Inasmuch as only a small part of the mucous membrane of the nose, namely, the so-called olfactory area, is supplied by the olfactory nerve, and the remainder, which is by much the larger part, by the fifth nerve, it is necessary to separate substances used for testing the sense of smell into those which stimulate the olfactory nerve and those which affect only the fifth nerve. If sensations of smell are blended with those of taste proper, such as bitter, sweet, salt, sour, a mixed sensation, termed a flavor, is produced. If the sense of smell be lost, the perception of flavors is also lost, but in spite of this fact, volatile substances, such as ammonia, acetic acid, or mustard, may so stimulate the nasal mucous membrane supplied by the fifth nerve that a distinct perception is aroused. Lachrymation may thereby be produced, due wholly to the irritation of nasal mucous membrane through the mediation of the fifth nerve. This, however, is not a true sense of smell. Bearing these facts in mind, tests for smell must be made with non-irritating substances, such as peppermint, oil of cloves, or asafetida, rather than with substances which irritate the mucous membrane.

**Diagnosis.**—After determining the fact of loss of smell by proper tests, it is essential to exclude disease of the mucous membrane, a purulent discharge, or other local cause for the defect. The retention of the ability to recognize flavors, with a loss of the sense of smell, is very frequently due to such lesions, owing no doubt to the fact that the expiratory stream is stronger and impinges more directly upon the olfactory area than the inspiratory stream. The importance of a local examination cannot be overestimated. In the examination, apart from the tests to which allusion has already been made, electricity has been locally applied. This procedure gives rise to a phosphorus-like odor, but the test is painful and at best of doubtful value. In making a routine examination account must be taken of the intelligence of the patient, and one nostril should always be closed when the other side of the nose is being examined. As a further complication it should not be forgotten that disease of the fifth nerve may at times cause considerable anosmia,



probably indirectly through the general nutrition of the mucous membrane, including the olfactory area.

**Prognosis.**—The outcome of affections of smell naturally depends essentially upon the character, persistence, and possibility of removal of the cause. Temporary loss of the sense of smell, due to local irritation of the nasal mucous membrane, as, for example, in ordinary rhinitis, is a matter of common experience. The function of smell is restored with the removal of the irritation. In man the sense of smell may be regarded as a luxury rather than a necessity. Hence its permanent loss, especially if unassociated with the loss of appreciation of flavors, is often not deeply felt, and in some cases no doubt not appreciated by the patient. Apart from injuries and infections of the nervous system the prognosis and treatment depend on the local conditions within the nasal cavity.

### OPTIC (SECOND) NERVE AND TRACT.

The optic nerve is to be regarded rather as an outgrowth of the brain than as a nerve properly so-called. As in the brain and cord, the axones running in the optic nerve are myelinated but devoid of a connective-tissue sheath. As in other sensory nerves, the primary neurones originate for the most part, in ganglion cells of the retina. Passing backward toward the brain a large number of fibres, presumably twice as many as uncrossed, cross at the chiasm. This crossing is partial in man, monkeys, and many mammals, and complete in fishes, amphibians, reptiles, and birds. A possible explanation for this irregularity of crossing is to be found in the fact that those animals whose vision is wholly monocular have a complete crossing, whereas those in which binocular vision has been established have developed a partial crossing. Behind the chiasm, the optic tracts, made up of an external fasciculus uncrossed, a middle fasciculus crossed and an internal fasciculus (Gudden's commissure) extend to their temporary termination in the lateral geniculate bodies, the pulvinars of the thalami and the superior corpora quadrigemina, constituting neurones of the first order. The inferior corpora quadrigemina are not to be regarded as a part of the visual system. The macular bundle presumably has bilateral cerebral connections, as shown by the fact of the frequent preservation of central vision in hemianopsia. The internal fasciculus of the optic tracts, known as Gudden's commissure, connects the two internal geniculate bodies, and is not to be regarded as immediately concerned with the visual act, except through possible reflex relations. From the dorsal portions of the thalami the external geniculate bodies and the superior corpora quadrigemina fibres pass through a secondary neurone system to the cortex of the occipital lobes in the region of the cuneus, constituting the so-called optic radiations. The visual representation in the cortex is considerably greater on the mesial aspect of the hemisphere than externally.<sup>1</sup> There is evidence to

<sup>1</sup> Much work has been done on the cortical localization of the visual centres. For summaries see Spiller, *The Cortical Centres of Vision*, p. 8, and Mills, *Position and the Subdivisions of the Primary Cortical Visual Area*, p. 75, in *The Eye and Nervous System*, Posey and Spiller, Lippincott, 1906. The exhaustive treatise by Wilbrand and Saenger, *Die Neurologie des Auges*, Wiesbaden, 1904, should be consulted in all matters relating to the ocular nerves.

show that certain fibres, originating in the cortex, pass to the retina through the superior corpora quadrigemina. A lymph space, known as the supravaginal space, presumably of importance in its relation to the results of intracranial pressure, surrounds the optic nerve and its sheath. The intervaginal space of the nerve communicates directly with the intracranial meninges (Schwalbe).

FIG. 19

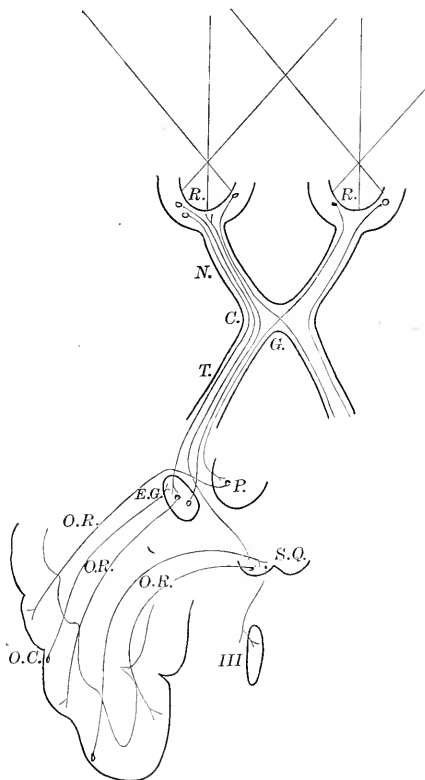


Diagram of the optic system. *R.*, retina; *N.*, optic nerve; *C.*, optic chiasm; *G.*, Gudden's commissure; *T.*, optic tract; *P.*, pulvinar of thalamus; *E.G.*, external geniculate body; *S.Q.*, superior quadrigeminum, the three ganglia constituting the primary visual centres; *O.R.*, optic radiations; *O.C.*, occipital cortex; *III*, nucleus of third nerve. Note crossing of internal fibres at the chiasm; also that most of the fibres originate in the retina, a lesser number in the cortex.

**Lesions of the Retina.**—An ophthalmoscopic examination often throws light on general diseases, and may be a most important element in diagnosis. Such an examination should not be omitted in ordinary routine examinations.

**Retinitis.**—Alterations of the retina, usually manifested as an inflammation, occur in a large number of conditions, a few of which only may with propriety be considered briefly here. The general characteristics of retinitis, as seen by the aid of the ophthalmoscope, are diffuse cloudiness, particularly of the more central portions of the fundus, due to a loss of transparency of the retina and veiling of the choroid. The optic papilla

shows more or less congestion with loss of clearness of outline, leading to a somewhat striated appearance. The veins are enlarged, tortuous, and engorged. Hemorrhages may occur with exudation, ultimately leading to a whitish appearance of the parts involved, through secondary changes. The color of hemorrhages, whether bright red, dark, or almost black, depends upon their age. If the nerve head or the choroid is involved in addition, the condition becomes a neuroretinitis or choroidoretinitis. Inasmuch as a retinitis is usually due to a constitutional infection, both eyes are commonly involved. The combination of the foregoing signs, together with certain characteristic groupings, make possible the recognition of somewhat definite forms of retinitis.

(a) **SYPHILITIC RETINITIS** (or syphilitic choroidoretinitis).—Syphilis manifests itself by somewhat varied lesions in the retina. The retinitis is usually associated with concurrent disease of the uveal tract, with frequent involvement of the choroid. The affection may be divided into two main types, one diffuse and the other circumscribed. In the diffuse type there is widespread but slight opacity of the retina with the occasional appearance of small grayish spots. The gradual disappearance of this opacity is accompanied by progressive changes in the pigment epithelium. The pigment cells occasionally wander into the retinal layers. In the circumscribed form, on the other hand, a characteristic feature is a yellowish-white exudate, occurring either in the macular region or along one of the larger vessels. When this occurs the ophthalmoscope will often disclose disease of the vessel wall. In the course of time these exudates are replaced by scar tissue, which predisposes to retinal detachment. Hereditary syphilis may likewise lead to a retinitis which is ordinarily observed only in the atrophic stage. There may be either small whitish areas, pigment clumps, or the connective-tissue remains of former exudates. In late cases vision is affected, and various scotomata and defects appear in the visual field. Night blindness is a somewhat constant feature, and occasionally micropsia (apparent diminution in the size of objects) and metamorphopsia (distortion of the outlines of objects) may occur. The treatment is of the underlying cause, syphilis.

(b) **ALBUMINURIC RETINITIS**.—The ophthalmoscope discloses hyperemia and swelling of the papilla and of the surrounding retina. Hemorrhages may occur in the nerve head and in the nerve-fibre layer of the retina. Irregular white patches may often be seen in the retina at a considerable distance from the papilla. The macula is not infrequently surrounded by a star-like arrangement of white dots, apparently converging toward it. An accompanying papillitis may be extreme, even suggesting the existence of a brain tumor. The white areas, the result of fatty degeneration, at times become confluent, forming single large white plaques. The hemorrhages vary widely both in size and number. An affection of the fundus occurs in albuminuric conditions, which is entirely uncharacteristic. For example, simple retinal hemorrhages, retinitis hemorrhagica, or even optic neuritis and choked disk may occur.

Retinitis may occur in anemia, leukemia, and many other general and constitutional diseases which come rather within the province of the ophthalmologist than the practitioner of internal medicine.

**Lesions of the Optic Nerve.**—Inflammation, in general limited to the optic disk, producing a so-called optic neuritis or papillitis, or, in extreme degrees of swelling, choked disk, is important to recognize. Although this sign may occur in a great variety of conditions, its usual cause is increased intracranial pressure from disease within the cranial cavity. Of such causes the most frequent are tumors, the location of which, rather than their character, is important in the production of a neuritis. Optic neuritis may also occur in other structural diseases of the brain, such as meningitis and abscess (rarely according to Bordley and Cushing) idiopathic hydrocephalus, and syphilis. In these conditions, in the absence of marked pressure, the mechanism of the production of the neuritis is often not easy to determine. Many cases of inflammation of the nerve head have been described in influenza, malaria, erysipelas, scarlet fever, and other general infections. It has also been observed occasionally in neuritis, anemia, and in intoxications from alcohol and lead. Certain deformities of the skull, and particularly localized disease of the orbital region, more or less directly involving the nerves, may naturally lead to their inflammation.

**Mechanism of Papillitis, or Choked Disk.**—Various theories, no one of which is entirely satisfactory, have been suggested to account for the so-called inflammation of the nerve head. Von Graefe held to the view that the condition was due to a venous stasis, resulting from an obstruction to the return flow of blood from the cavernous sinus (*Stauungspapille*). On the basis of Schwalbe's work on the communication between the sheaths of the optic nerve and meninges, Manz and Schmidt-Rimpler concluded that mechanical pressure led to an œdema of the disk. Meehan regards as the important elements, œdema, congestion, and inflammation, due to a dropsy of the optic nerve lymph space induced by increased intracranial pressure. Parinaud considers choking of the nerve head to be due to an œdema of the optic nerve itself. A somewhat widely accepted theory, of which von Leber is an advocate, bases the optic neuritis upon the supposed presence of an irritant in the fluid distending the nerve sheath, produced in some way by the presence of a tumor or other source of irritation within the cranial cavity. A significant investigation by Bordley and Cushing<sup>1</sup> tends strongly to support the so-called mechanical theories. Of the various terms used to describe the condition of the nerve they prefer "choked disk" to designate all grades of the neuroretinal œdema. The view expressed is that early injection with stasis of the retinal vessels, œdema with elevation of the papilla, and final cellular infiltration with new tissue-formation leading to atrophy are stages of the same process, due essentially to the distension of the sheath of Schwalbe by obstructed cerebrospinal fluid. It will be seen that the toxic element plays small part in this conception. The results upon the optic disk of decompressive cranial operations lead to the assumption that choked disk from practically all causes, including the changes occurring in nephritis, are due to mechanical rather than toxic causes. The evidence in this article is strongly in favor of the mechanical theory. It must,

<sup>1</sup> *Jour. Am. Med. Assn.*, 1909, lii, 353.

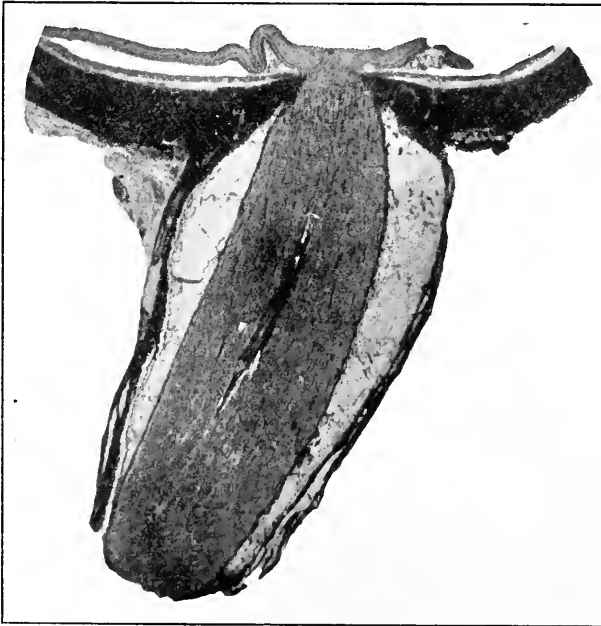
however, still be regarded as possible that different causes may lead to similar ophthalmoscopic appearances. A true neuritis behind the bulb

FIG. 20



Optic neuritis (choked disk). From a case of orbital tumor. Very slight œdema of subvaginal lymph space. (Reproduced through the courtesy of Dr. Frederick H. Verhoeff.)

FIG 21



Edema of subvaginal lymph space of optic nerve. With slight swelling of the disk, from a case of tuberculous meningitis. (Reproduced through the courtesy of Dr. Frederick H. Verhoeff.)

has also been observed in certain cases, and an engorgement œdema is a somewhat characteristic accompaniment of the typical picture of choked

disk. If, on the other hand, a simple inflammation is the predominant factor, the elevation of the disk is less marked, and the retina is more apt to be involved. A marked swelling of the nerve head is in general to be regarded as an indication of pressure behind. The term optic neuritis is rather an unfortunate one. Papillitis more nearly describes the condition, but both terms are bad if it can be shown that inflammation plays no part. The term choked disk is useful as indicating a characteristic ophthalmoscopic appearance, associated with much swelling, but it may properly be given a much wider significance.

**Appearance of the Optic Disk in Papillitis.**—A somewhat artificial distinction may be made between a first stage of congestion with œdema and a second stage of true neuritis or papillitis. The general appearances are as follows: The nerve head early shows a deepening of color, with a gradual loss of outline. As the process goes on the optic disk swells, with a final complete loss of its form, so that ultimately its position can only definitely be determined by the convergence of bloodvessels toward its centre. The height of the swelling is often several millimeters. The appearance of the vessels is perhaps most characteristic. The arteries are small, and frequently concealed by swelling; the veins are dark, distended, tortuous, and appear divided, owing to infiltration of the disk. Small flame-like hemorrhages are frequent, but not an essential part of the picture. Vision may, for a long period, be unaffected, and the optic neuritis, therefore, go unnoticed. The visual fields show an irregular concentric contraction. There is often an increase in the size of the normal blind spot and loss of appreciation of red and green. In brain tumors, particularly, such a papillitis may become what has been well described as a choked disk. A condition known as descending neuritis is associated with moderate swelling, and considerable exudation, producing discoloration and opacity of the papilla, but not limited to it. The retina may likewise be involved, leading to a neuroretinitis.

**Optic Atrophy.**—A degeneration of the optic nerve may occur either as a primary process or as secondary or consecutive to a papillitis. The general appearances of atrophy of the optic nerve when well marked are as follows: Alteration in color, varying from gray to greenish-gray to clear white. Slight variations in color are extremely difficult to determine, and their significance should not be ventured except by those of wide experience in ophthalmoscopy. There is sinking of the centre of the disk, often with a characteristic appearance of the lamina cribrosa. The margins of the disk are usually distinct, but in secondary atrophy following papillitis they may be veiled for a long period. A broadening of the scleral ring indicates a shrinkage of the disk. The arteries, particularly in consecutive atrophies, are narrowed, in contrast with the veins which often retain their tortuosity. There is, however, finally an atrophy of the veins as well, with secondary connective-tissue changes. It is evident that the transition from papillitis to atrophy is extremely gradual, and that the foregoing appearances are clearly defined only when the atrophy has progressed to a considerable degree.

In contrast to papillitis, vision always suffers in optic atrophy. There are various defects in the field and development of central scotomata with

concentric limitation. The loss of the color field is usually greater than that of the form field. The pupillary light reflex suffers in proportion to the degree of atrophy.

*Primary atrophy* of the optic nerve occurs in tabes dorsalis, paralytic dementia, and, much less commonly, in multiple sclerosis. In the latter disease a pallor of the temporal half of the disk, as pointed out many years ago by Uhthoff, is not an uncommon appearance, presumably induced by partial lesions of the chiasm. Primary atrophy may likewise occur in chronic malaria, diabetes, syphilis, arteriosclerosis, and through the toxic action of certain drugs. A curious and inexplicable atrophy of the optic nerve on a hereditary basis has been described, usually appearing between the eighteenth and twenty-third years. A stage of œdema and congestion, gray discoloration and pronounced atrophy, may be distinguished in this affection.

*Secondary atrophy* is usually consecutive to an optic neuritis, but may occur through external causes leading to compression of the tracts and fibres of the nerve. The pathological appearances of optic nerve atrophy are similar to those of the degeneration of fibres in general.

*Diagnosis.*—Although an optic neuritis may often be a determining point in diagnosis, its occurrence in unexpected conditions and its not infrequent failure to develop in well-defined, intracranial disease should lead to hesitation in its interpretation. Lesions of the spinal cord, for example, may occasionally give rise to a well-defined optic neuritis. Weisenburg and Thorington<sup>1</sup> reported a definite optic neuritis in a case of syringomyelia, and the writer has observed the development of a neuritis, rapidly leading to blindness, in a case of primary tumor of the lower portion of the cord extending upward.<sup>2</sup> In both these instances the optic neuritis is, no doubt, to be attributed to a blocking back of cerebrospinal fluid leading to hydrocephalus. Optic neuritis occurring in the course of a myelitis has been described, and presumably is to be attributed to the action of the toxic agent leading to the cord disease.<sup>3</sup> On the other hand, cases of brain tumor are in a considerable proportion of cases unaccompanied by optic neuritis, and especially in those in which pressure is not a marked symptom. The occurrence of optic neuritis in the course of intracranial syphilis is not infrequent, particularly in those cases in which the symptoms develop in acute fashion. Cases might easily be multiplied to illustrate the uncertainty of optic neuritis in various serious intracranial conditions. The fact remains that the ease of its demonstration, and its definite significance when present make a study of the eye-grounds in all cases a matter of the utmost importance.

*Course and Prognosis.*—The course and outcome of affections of the optic disk are wholly dependent upon the character of the cause, whether it acts temporarily or permanently and whether it is capable of removal.

**Retrobulbar Neuritis.**—An acute form of retrobulbar neuritis has been described, among other causes, in influenza, multiple sclerosis, and poisoning by wood alcohol. In this condition there is rapid loss of sight in one

<sup>1</sup> *Am. Jour. Med. Sc.*, 1905, cxxx, 1019.

<sup>2</sup> Taylor and Collier, *Brain*, 1901, xxiv, 532, discussed this in detail.

<sup>3</sup> Kernhensteiner, *Münch. med. Woch.*, 1906, liii, 802.

eye, occasionally in both, with the development of a central scotoma and pain in the orbit. At first there are no changes in the optic nerve; later an optic neuritis may appear passing into atrophy, or the atrophy may apparently be primary. Complete blindness is rare, but a defective vision may remain. A chronic form of retrobulbar neuritis is more frequent. This also is rapid in onset; objects are veiled; the acuteness of vision is reduced; there is loss of color sense, with the development of a central scotoma, particularly for colors, usually oval in form. In extreme cases a central amblyopia may develop with complete loss of central vision. This is usually binocular, and often unobserved by the patient. Recovery is partial or complete. The special causes are excess in alcohol, tobacco, or both, hence it is more frequent in men than in women. The ophthalmoscopic appearances are hyperemia of the papilla, loss of outline, with the development of white striæ along the vessels. Following this temporary phase there is apt to be a pallor of the temporal side of the disk, the nasal portion remaining normal. Rarely the whole papilla undergoes a so-called white atrophy. The essential treatment is total abstinence from alcohol, tobacco, or other causal agents.

**Amblyopia and Amaurosis without Ophthalmoscopic Changes.**—A great variety of conditions affecting sight may occur entirely without objective evidence in the fundus. Some of the more important of these are as follows: A congenital disturbance of vision for colors, or total color blindness; an amaurosis due to trauma, uremia, diabetes, malaria, from loss of blood, from the abuse of drugs, of which methyl or so-called wood alcohol may be taken as an example; various disturbances of vision in hysteria with inversion of color field; monocular diplopia; conjugate deviation and general narrowing of the visual fields are the more important. Night blindness (nyctalopia), day blindness (hemeralopia), red vision (erythropsia), reduced size of objects (myopsia), and increased size of objects (megalopsia) are some of these disturbances of vision.

**Vision.**—Apart from the ordinary tests by means of charts, it is often useful and essential to diagnosis to determine the degree of eccentric vision and its possible limitation. Such an examination may be made roughly by means of the hand or any other object brought into the field of vision from various directions. Gross defects may be determined by carefully observing the point at which the patient first sees the object. Hemianopsia may easily be so determined. A somewhat more accurate method is to require the patient to look at a definite mark on a blackboard while seated at a distance of from one and a half to two feet. An object, as, for example, a piece of chalk, brought into the fields from various points, when recognized by the patient as a moving object, serves as an indication of the extent of the visual field in that direction, and may be so indicated on the board. In making such examinations one eye should be closed while the other is being examined. Colors are less easily seen in the order from without inward, blue, green, red. A perimetric examination is essential for an accurate determination of the visual fields.

**Disturbances of Vision.**—The general disturbances are: (1). Concentric narrowing of the fields; (2) scotomata, or defects of vision elsewhere than



on the periphery; (3) hemianopsia. Mention has been made of concentric narrowing and various scotomata. Hemianopsia is a disturbance of more importance for our present purpose, inasmuch as it indicates not only disturbance in peripheral vision, but also is of very great importance in determining the location of lesions within the cranial cavity.

**Affections of Optic Chiasm and Tracts.—Hemianopsia.**—Destructive lesions of the optic nerves, ventral to the chiasm, naturally give rise to

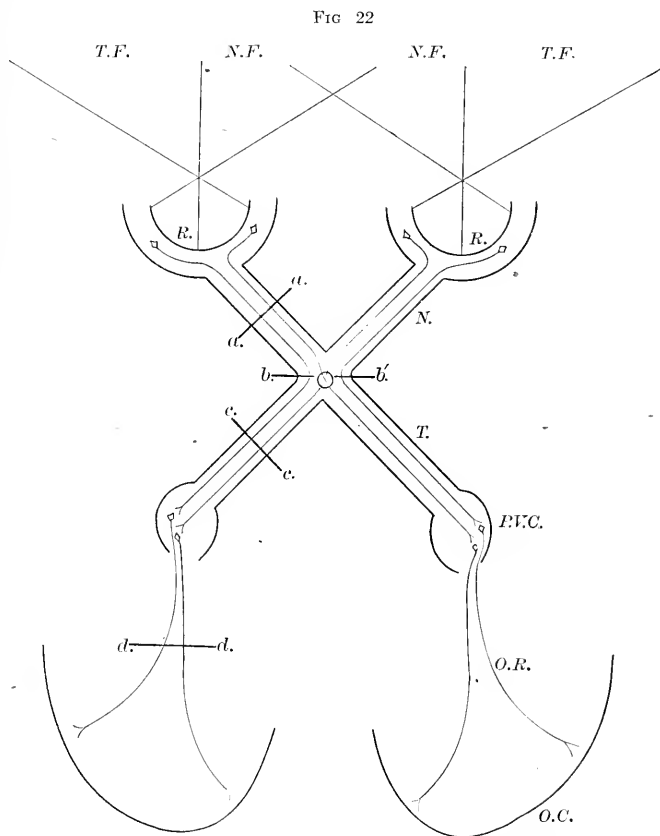


Diagram of lesions producing hemianopsia. *T.F.*, temporal field; *N.F.*, nasal field; *R.*, retina; *N.*, optic nerve; *T.*, optic tract; *P.V.C.*, primary visual centres; *O.R.*, optic radiations; *O.C.*, occipital cortex; *a.a.*, lesion producing unilateral blindness; *b.*, lesion producing unilateral hemianopsia; *b.b'*, lesions producing binasal hemianopsia, very rare; *O.*, at centre of chiasm, lesion producing bitemporal hemianopsia; *c.c.*, *d.d.*, lesions producing homonymous hemianopsia. Hemianopic pupillary reflex in lesions anterior to primary visual centres, and behind the chiasm.

unilateral or bilateral blindness and need no further mention. Owing to the partial crossing in the chiasm, however, partial disturbance of vision develops when the lesion lies either at that point or in the course of the optic tracts to their central terminations, characterized by losses of portions of the visual field in each eye. The usual lesion of the chiasm occurs in its central portion, leading to disturbance in the nasal portions of the retina with consequent affection of vision in the temporal field.

This condition is known as bitemporal hemianopsia. Naturally, if the process in the chiasm extends, total blindness may result, or total blindness in one eye with retained half vision in the other. The occurrence of binasal hemianopsia, through symmetrical lesions limited to the outer chiasm bundles, is exceedingly unusual, as well as lesions of the chiasm producing inferior and superior hemianopsia. Syphilis, tumors, particularly of the hypophysis, and hydrocephalus are the more frequent causes of disturbances in the chiasm leading to hemianopsia.

Affections of the optic tract lead to homonymous bilateral hemianopsia, due to the fact that each optic tract contains fibres supplying the temporal side of one retina and the nasal side of the other, the fibres to the nasal side being crossed, and to the temporal side being uncrossed. The same visual disturbance is produced by any lesion either of the primary optic centres or of the occipital lobe of one side. With the exception of the so-called Wernicke hemianopic pupillary reaction the localizing significance of homonymous hemianopsia is relatively slight, apart from coincident lesions of neighboring portions of the brain or cranial nerves. Affections of the visual area in the occipital cortex, produced by tumor, areas of softening or injuries, may lead to isolated hemianopsia. The same is true of lesions of the optic radiations, which, however, if affected on the left side, usually are accompanied by aphasia. Certain facts of importance regarding the location of the lesion producing the hemianopsia may be derived from a detailed study of the visual field, particularly with reference to the dividing line between the seeing and the blind areas. In complete lesions of the optic tract the hemianopsia is usually complete, by which is meant that it passes vertically through the fixation point. Such a complete hemianopsia is less frequent in disease of the external geniculate body, and occurs only exceptionally in affections of the cortex. The incompleteness of the hemianopsia in lesions above the optic tracts must be regarded as due to the separation of the fibres and the probable separate representations of the visual fields in the cerebral cortex. The frequent sparing of the macula, for example, may be explained on the basis of a separate cortical localization for that group of fibres.

The association of homonymous hemianopsia with concentric narrowing of the visual field is, no doubt, often to be explained, as Uhthoff has pointed out, by a complication of organic disease with a functional disturbance. Affections of the color fields, appearing in hemianopic form, have been described, and various minor anomalies of the more definite picture which is here presented. Bilateral hemianopsia is essentially identical with blindness, except that in several cases central vision has not been destroyed, and the light reflex of the pupil is retained, provided the lesion lies beyond the reflex arc.

**Hemianopic Pupillary Reaction.**—A further possibility of localizing the lesion producing hemianopsia is afforded by the so-called hemianopic pupillary reaction, to which Wernicke has called attention. If the lesion lies within the reflex arc from the retina to the central visual centres, illumination of the amblyopic half of the retina will lead to no pupillary reaction, owing to the severance of the reflex arc. Such illumination

applied to the seeing half of the retina will produce the ordinary reaction. If, however, the lesion lies behind the reflex arc between the optical centres and the visual cortex, illumination of the blind side of the retina will produce a normal pupillary response. This test is of value, but its successful performance demands much care, and the isolated illumination of one retinal half is difficult of accomplishment.

**Mind Blindness.**—In the consideration of the disturbances of the optic nerve the condition of so-called mind blindness demands mention. This state is not to be confused with optical aphasia. In this latter condition objects are both seen and their significance recognized, but cannot be designated in words through the medium of the sense of sight. In such cases the proper designation may, at times, be aroused through another sense, as, for example, a key may be recognized as a key, but cannot be named until it is put into the patient's hand and a demonstration of its use requested.

### OCULOMOTOR, TROCHLEAR, ABDUCENS (THIRD, FOURTH, SIXTH) NERVES.

The three nerves, third (oculomotor), fourth (trochlear), and sixth (abducens), which control the movements of the eyeball and pupil, may be best grouped, although the function of each nerve is distinct and demands individual consideration. The relations of the three nerves governing the movements of the eyeball are extremely close, which naturally is a development of the function of binocular vision. It is also of interest and of very great clinical importance to recognize the connection between these three motor nerves and the sensory eighth nerve. The preservation of equilibrium is undoubtedly dependent upon this relationship, with the complex reflex connections of the eighth nerve with the cerebellum and other portions of the central nervous system.

**Symptoms of Paralysis of Ocular Nerves.**—A complete paralysis of all the muscles providing for the movements of the eye gives rise to the condition known as total *ophthalmoplegia*. This may develop partially either as an internal ophthalmoplegia, in which the pupillary muscles are chiefly affected, or as an external ophthalmoplegia, in which those governing the globe are alone involved.

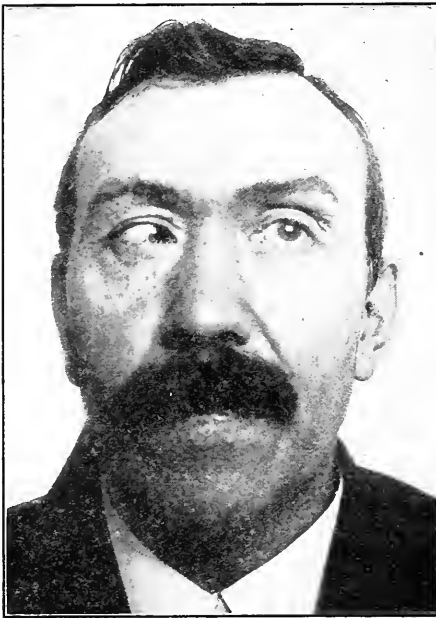
Under normal conditions the external rectus muscle (sixth nerve) moves the bulb directly outward. The internal rectus (third nerve) moves it directly inward; the superior rectus (third nerve) upward and inward, also with rotation of the upper axis slightly inward; the inferior rectus (third nerve) draws the globe downward and slightly inward. The superior oblique (fourth nerve) moves it downward and outward with inward rotation of the upper end of the vertical axis. The inferior oblique finally draws it upward and outward, with outward rotation of the upper end of the vertical axis.

A defect of the lateral action of the eyes, normally brought about by the internal rectus of one eye and the external rectus of the other, is usually easily observed. If the movement of both bulbs to one or the

other side is interfered with, the condition is known as conjugate deviation or associated paralysis (*Blicklähmung*). The paralysis of eye muscles leads to the following conditions: (1) Loss or limitation of special movements of the bulb; (2) diplopia; (3) secondary contracture of the antagonists; (4) secondary deviation of the sound eye; (5) false projection of the visual field and abnormal position of the head.

From the general clinical standpoint the development of double vision is particularly important as an indication of more or less serious intracranial disease. It should be borne in mind that in congenital conditions or in persons in whom a strabismus has existed for a long time, a diplopia must exist, although often not recognized by the patient.

FIG. 23



Paralysis of left sixth nerve. Occurring in the course of tabes. (Massachusetts General Hospital.)

This is due to the fact that through training and habit the retinal image of one eye is ignored, after the manner of a person looking through a microscope with both eyes open. On the other hand, in a diplopia coming on suddenly the double images are at first annoying to the patient, often leading to the semivoluntary closure of one eye to overcome the confusion of a double image. It is well to remember this in arriving at a conclusion regarding the length of time a diplopia has existed. A simple examination, asking the patient to fix the moving finger of the examiner with both eyes, is often enough to determine the existence of double vision. The patient should be directed to state the exact moment at which two pictures come into the field. It is sometimes useful to use a colored glass before one eye in order that the two images may be sharply distinguished. The image seen by the sound eye is spoken of as the

true image, that by the affected eye as the false image. If the false image is on the same side as the eye by which it is seen, the diplopia is spoken of as homonymous, otherwise it is spoken of as crossed. If, for example, the right image disappears with the closure of the right eye, the diplopia is homonymous. In general, convergent strabismus is accompanied by homonymous, divergent strabismus by crossed diplopia. The position of the head is often altered in order to overcome the annoyance produced by a diplopia. Such altered position of the head is strikingly seen in cases of conjugate deviation, in which it is impossible to bring the eyeballs into the median line. The curious condition of so-called monocular diplopia

or polyopia is of rare occurrence. It has been observed and adequately described in hysteria (Janet, Prince).

**Paralysis of Special Muscles.—External Rectus.**—This muscle is frequently paralyzed through lesion of the sixth nerve at some part of its long course from the pons forward. A convergent squint is produced by this paralysis, which is increased through secondary contracture of the internal rectus. The diplopia is homonymous, with a widening of the space between the upright images as the object is moved outward toward the side of the paralyzed muscle.

**Internal Rectus.**—Isolated paralysis of this muscle gives rise to a defect of inward movement with consequent divergent squint. There is a crossed visual image in that part of the field corresponding to the unaffected eye. The space between the images increases as the object is moved toward the side of the sound eye.

**Superior Rectus.**—Limitation of upward movements with rotation of the bulb, due to the tension of the inferior oblique, is induced by paralysis of this muscle. There is a crossed double image in the upper half of the field. The upper or false image is oblique and separates from the true image on looking upward.

**Inferior Rectus.**—Paralysis produces downward limitation. The eye is turned outward and also is rotated outward in the effort to look down (superior oblique). The diplopia is crossed in the lower half of the field. The false image is lower than the true and the distance between them increases downward. The images are oblique.

**Inferior Oblique.**—Movements upward and outward are limited. On looking upward the bulb is rotated inward. There is homonymous diplopia in the upper half of the field. The images are oblique.

**Superior Oblique.**—Paralysis of this muscle produces slight disturbance of motion. On looking downward there is a slight turning inward of the eye, with homonymous diplopia in the lower half of the field and convergent strabismus. In this paralysis on going down stairs the steps often appear to be double.

A slight *exophthalmos* is produced by a paralysis of all the external muscles of the eye, which, on account of the immobility of the globe, is not likely to be confounded with the *exophthalmos* of Graves' disease, hydrocephalus, or postorbital tumors. The paralyzes of convergence and divergence which occasionally occur have been supposed to be due to disturbance in a special cortical centre which presides over these associated movements. Such paralyzes are apt to be accompanied by vertigo. Conjugate deviation of the eyes, due either to spasm or paralysis, is a sign of considerable diagnostic importance and is strongly indicative of a lesion within the pons having relation to the dorsal longitudinal bundle through which the associated movements of the third and sixth nerves are brought about. Spiller<sup>1</sup> has discussed this matter in detail, and reaches the following conclusions: Persisting paralysis of associated lateral movement indicates a lesion of the posterior longitudinal bundle. Persistent paralysis of associated upward or downward movement

<sup>1</sup> *Jour. of Nerv. and Ment. Dis.*, 1905, xxxii, 417.

indicates a lesion in the vicinity of the oculomotor nucleus. Such paralysees are not the result of a lesion of extracerebral nerve fibres. Cortical lesions may produce such paralysees, but they are temporary unless due to a bilateral brain lesion. In hysterical states such associated paralysees may likewise occur and also as a result of inflammatory lesions or others produced by alcohol and syphilis; tumor is a common organic cause, and if such associated paralysees persist operation can never be regarded as more than palliative.

*Nystagmus* is commonly met with in conditions of muscular inequality, in multiple sclerosis, and as one of the signs of lesions in the posterior fossa, particularly in relation to the eighth nerve. Although the cause may be varied, the general explanation must be sought in a loss of coördination between the various muscles of the eye. Depending upon the movement which the eyeball makes, the nystagmus is spoken of as horizontal, vertical, or, more rarely, rotatory. The movements are very much more marked on lateral fixation, and it should be borne in mind that under these conditions considerable oscillation of the bulbs must be regarded as within normal limits. Nystagmus is often conspicuous in the blind, with whom fixation is not possible, and also in persons, especially children, with defective vision.

**Intra-ocular Muscles.**—The muscles of the iris, associated with alteration in the size of the pupil, are the so-called constrictor and dilator pupillæ. The ciliary muscle, presumably important in bringing about the act of accommodation, consists of unstriped longitudinal and circular fibres forming a circular band about one-eighth of an inch in breadth between the choroid and the iris. The nerve of these muscles is through the nasal branch of the ophthalmic division of the third nerve and the short ciliary nerve from the ciliary ganglion. The circular fibres of the iris receive their innervation from the third nerve and the radiating fibres from the sympathetic, hence paralysis of the third nerve leads to dilatation of the pupil, and paralysis of the sympathetic to its constriction. Naturally the opposite results occur when the lesion of the nerves involved is irritative rather than destructive.

The pupils narrow physiologically under the following conditions: Light on the retina; consensual reaction or the phenomenon of pupillary constriction of one eye when light is thrown into the other; narrowing of the pupil for accommodation for near objects. The path by which the reflex action of the pupil is determined is still in dispute. The so-called reflex centre has been variously placed in the ciliary ganglion, in the oculomotor nucleus, the corpora quadrigemina, the upper portion of the spinal cord, and in the oblongata. Experimental and clinical studies give no uniformity of result as to the exact location of this reflex centre. Spiller is inclined to think that there is a limited area in the oblongata near the respiratory centre which is inhibitory in function, that cerebral to this there is a subordinate centre, and that the complete reflex is, therefore, conditioned by one centre having its location in the corpora quadrigemina and another at the spinal end of the fourth ventricle. It is probable that the optic nerve contains special fibres which are not identical with those fibres which subserve sight, but which con-

stitute a part of the reflex arc partially crossing in the chiasm. In rare instances these fibres have apparently retained their reflex function when the optic nerves have been extensively diseased.

**Pathological Variations in the Pupil.**—The ordinary pathological alterations may be summarized as follows, after Uhthoff, quoted by Weeks: (1) Loss of pupillary light reflex with retained convergence and accommodation (Argyll-Robertson pupil); (2) loss of convergence and accommodation with retained light reaction; (3) loss of pupil reflex for light and accommodation; (4) loss of all reflex movements of the pupil (ophthalmoplegia interna); (5) loss of direct light reflex in an amaurotic eye and consensual light reflex in the fellow eye due to loss of vision from a lesion peripheral to the chiasm; (6) loss of sensory or psychical reflex action; (7) abnormal miosis with retention of light and convergent reflexes arising from stimulation of the sphincter or paralysis of the dilatator; (8) abnormal dilatation of the pupil (spastic mydriasis) due to stimulation of the dilatator or paralysis of the sphincter with retention of light and convergent reflexes; (9) difference in size of the pupils (anisocoria); (10) changing anisocoria; (11) irregularity of pupil outline; (12) change in the size of the pupil under uniform illumination (hippus); (13) paradoxical pupillary reflex or dilatation where contraction would ordinarily be expected; (14) hemianopic pupillary reaction.

Of these conditions several only require special comment here. From a practical standpoint the Argyll-Robertson pupil is of very great importance. In testing for this reaction it should be remembered that in elderly persons a light reaction is naturally slow, that there are many transitional conditions between a normal pupillary reflex and an immobile pupil, and that an accompanying convergence or accommodative reaction must not be mistaken for a light reaction. Although opinions differ, syphilis is possibly not always the cause of this peculiar pupillary phenomenon. It is most often met with in the syphilitic disease, tabes. In 166 cases Uhthoff found an Argyll-Robertson pupil associated with tabes in 67.6 per cent., with dementia paralytica in 8.8 per cent., in syphilis without tabes in 8.17 per cent. In other cerebral diseases it occurred in only 5.87 per cent., and the remaining very small percentage in other disorders of the nervous system. In a statistical summary by Siemerling, made on the basis of 1639 cases of Argyll-Robertson pupil, 94.7 per cent. occurred in persons suffering from general paralysis, tabes, or syphilis.<sup>1</sup>

As previously stated, the location of the lesion producing this sign remains in doubt. Its frequent occurrence in tabes had led to the supposition that the cord was primarily responsible, but the evidence for this is insufficient. Internal ophthalmoplegia, due to the paralysis of the ciliary and sphincter muscles, is in many cases presumably dependent on a nuclear lesion, with syphilis often conspicuous in the etiology.

Slight differences in the size of the pupils are not to be regarded as pathological. If pronounced, however, suspicion should be aroused and attention directed toward the possibility of tabes, dementia paralytica, tumor of the neck, or tumor of the upper portion of the cord. Irregularity

<sup>1</sup> Bach, *Pupillenlehre*, Berlin, 1908, p. 144.

of pupillary outline, unless due to local disease of the eye, is to be attributed in most cases to lesions of the nerves innervating the iris. Such a condition of the pupils is often an early sign in tabes and dementia paralytica, and may precede the development of an Argyll-Robertson pupil.

**Special Conditions of Paralysis of Ocular Nerves.**—Owing to the wide distribution of the third nerve to both the external and internal muscles of the eye, its paralysis gives rise to the most striking signs. If all branches of this nerve are affected by the lesion, the paralysis induced gives rise to external squint, ptosis, dilated and inactive pupil, and certain other less conspicuous defects in the movement of the eye from involvement of other muscles than the internal rectus. Owing also to the wide distribution of the nerve, individual muscles are often the seat of paralysis, explained in some instances at least by the separation and distinct representation of the individual muscles in the nuclei of origin. Such partial palsies are, in fact, more often met with than the complete paralysis, and occur notably in tabes, basal gummata, or meningitis, and in other conditions within the cranium which lead to pressure or partial destruction of the nerve. Among the common more or less isolated paralyses of muscles supplied by the third nerve, ptosis takes the first place. A great variety of conditions may produce this disturbance. It may, for example, occur as a congenital condition in which there is from birth a certain incapacity to open the eye widely through weakness of the levator. This affection appears therefore to have a hereditary basis.<sup>1</sup> Acquired ptosis may arise from traumatism, vascular disorders, increase of intracranial pressure, and notably in myasthenia gravis and in Gerlier's disease. Ptosis is extremely frequent as part of the more general involvement of the third nerve. In relatively young persons this sign, often associated with paralyses of other ocular muscles, is significant of syphilitic infection. The sixth nerve supplying the external rectus is likewise frequently involved in paralytic conditions, giving rise to internal squint. An association of polyuria with paralysis of the sixth nerve has been described. The causes of its paralysis are, however, in general similar to those affecting the third nerve. Paralysis of the fourth nerve is often difficult to determine and is of small practical significance. It is rarely involved alone.

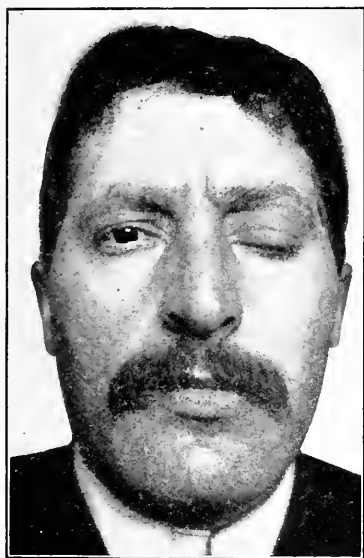
**Recurrent Oculomotor Palsy.**—This extraordinary condition, as its name implies, is a recurrent palsy of one third nerve and always the same nerve, occurring usually at regular intervals, lasting days, weeks, or months, and then disappearing in whole or in part. Children are usually affected. The paralysis is accompanied by pain in the head or through the eye on the side affected, also by nausea and vomiting, in this respect having a certain analogy to migraine. Unlike migraine, the headache and vomiting may continue for a week. Often there is headache more or less severe at intervals of about a month, at times accompanied by oculomotor paralysis. In typical cases the whole supply of the oculomotor nerve is affected; in others, individual muscles. A somewhat doubtful distinction has been made between those cases in which there is

<sup>1</sup> A family type of ptosis has been described (E. W. Taylor) occurring in persons over fifty, associated with progressive paralysis of deglutition.



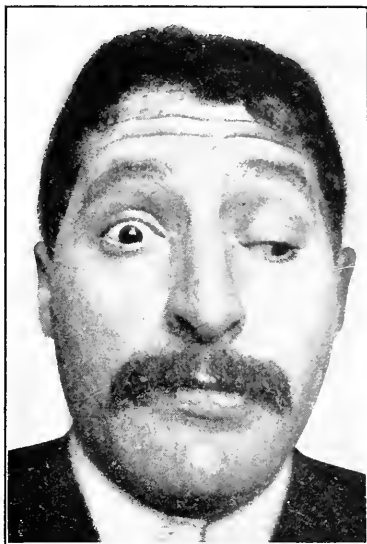
an interval entirely free from paralysis and those in which a certain amount of paresis persists. Oppenheim, following Charcot's theory,

FIG. 24



Paralysis of left third nerve. Case of ophthalmoplegic migraine. (Dr. G. A. Waterman.)

FIG. 25



Same as Fig. 24. Attempt to open eye. Showing action of frontal muscle. (Dr. G. A. Waterman.)

FIG. 26

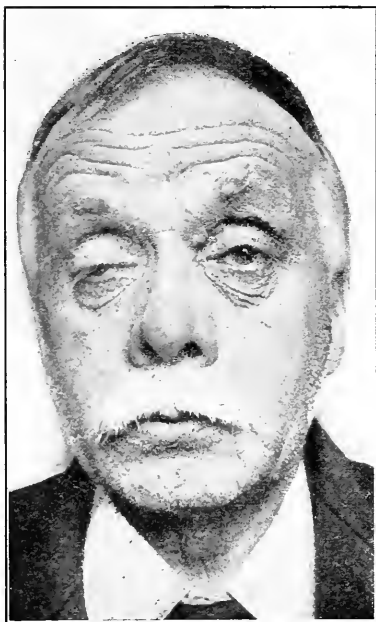


Double ptosis. An operation to raise the lid has been attempted on the right side. (Massachusetts General Hospital.)

finds the most probable explanation in a vasomotor disturbance allied to ordinary hemicrania. In general, the relations of the condition to ordinary ophthalmic migraine<sup>1</sup> and to the less frequent ophthalmoplegic migraine is a matter of very great interest, a discussion of which is not in place here. A similar periodic abducens and trochlear paralysis has been described. Some of the cases are apparently progressive, others stationary. The prognosis is doubtful, and the treatment uncertain, owing to the lack of exact knowledge of the predisposing causes.

**Ophthalmoplegia Interna.**—This, on the whole, unusual condition, is due to a paralysis of the sphincter pupillæ and ciliary muscles, and is usually unilateral. Rarely one of these muscles may be involved without the other, and not infrequently the process may extend to the extrinsic muscles of the eye, to involvement of other muscles supplied by the third nerve, as well as to those supplied by the fourth and fifth nerve. The lesion is presumably nuclear. Syphilis is apparently the most frequent cause, but such a paralysis has also been observed in diphtheria.

FIG. 27



External ophthalmoplegia. Involvement of both third nerves and of left sixth nerve. All movements of eyeballs imperfect except of the right outward. Normal pupillary reactions. (Massachusetts General Hospital.)

**Ophthalmoplegia Externa.** — More commonly a paralysis of the external eye muscles, with or without a coincident disturbance of the intrinsic muscles, occurs in the form, if the onset is acute, of a so-called superior encephalitis, or, if the course is from the outset progressive, as chronic ophthalmoplegia.

**Superior Encephalitis (Wernicke).** —In 1881 Wernicke<sup>2</sup> described a symptom complex which he called acute hemorrhagic superior poli-encephalitis. Alcohol was regarded as playing an important part in the etiology in two of the three cases originally described, and inflammatory changes were found in the neighborhood of the third nerve as the pre-

dominant lesion. The essential symptoms were paralysis of ocular muscles, of quick onset and rapid progress, leading finally to a practically total paralysis of the muscles involved, death resulting in from ten to fourteen days, with certain general cerebral disturbances suggestive of

<sup>1</sup> Walton has advanced the idea that ordinary migraine is an occupation neurosis, involving the visual centres, centre of accommodation, intrinsic and extrinsic muscles of the globe and certain muscles outside the orbit, *Jour. Am. Med. Assn.*, 1908, li, 200.

<sup>2</sup> *Lehrbuch der Gehirnkrankheiten*, 1881, Sec. 47, 229.

alcoholic intoxication. Further observation has shown that Wernicke's conception must be widely extended, and that a fairly well-defined group of conditions exists, due apparently to infection in the broad sense, which are characterized by a great variety of paralyses of ocular muscles, both external and internal. The pathological anatomy is not uniform, but various nuclear and peripheral degenerations have been found. There is increasing evidence to show that the relationship between this so-called superior encephalitis, poliomyelitis, and various other acute inflammatory conditions of the central nervous system is an exceedingly close one, if, in fact, this whole group of conditions is not identical.<sup>1</sup>

**Chronic Ophthalmoplegia.**—Although this condition is hardly to be regarded as a disease *sui generis*, its striking symptomatology justifies its separate consideration. In most cases this usually progressive form of paralysis of eye muscles forms part of a more general process, and is often associated with bulbopontine degenerations of motor nuclei and occasionally with degenerative cord lesions. The symptom-complex, originally described by von Graefe, is characterized by a slow development and long course, inasmuch as no centres essential to life are involved in its progress. The affection usually begins with involvement of a single muscle, often the levator palpebræ, followed by gradual extension to the external muscles of both eyes, often sparing the accommodative mechanism. The character of the paralysis points strongly toward nuclear degeneration as a cause. Finally, the eyeball may become entirely immovable, with involvement of the orbicularis oculi. The disease is usually progressive and ultimately fatal. The usual lesion found postmortem is a chronic nuclear degeneration entirely analogous to progressive bulbar palsy and progressive muscular atrophy.

### TRIGEMINAL (FIFTH) NERVE.

**Ophthalmic Branch.**—The first or ophthalmic branch supplies the eyeball and lachrymal glands, the conjunctiva, except that of the lower lid, the skin of the forehead and scalp up to the vertex, the mesial part of the skin of the nose and the mucous membrane of the upper part of the nasal cavity. There are also afferent pupil dilating fibres derived from the cervical sympathetic.

**Superior Maxillary Branch.**—The second, or superior maxillary division, is connected with Meckel's ganglion in the sphenomaxillary fossa which gives off the vidian nerve, important in relation to the sense of taste. The ultimate supply of the nerve is to the skin of the upper lip, side of the nose and adjacent part of the cheek, the lower eyelid, and part of the temple, also to the conjunctiva of the lower lid, the upper teeth, mucous membrane of the upper lip, upper jaw, uvula, tonsil, nasopharynx, the middle ear, and lower part of the nasal cavity.

**Inferior Maxillary Division.**—The third, or inferior maxillary division, so far as it is sensory, supplies the skin of the posterior side of the temple, adjacent part of the pinna, anterior and upper wall of the external auditory

<sup>1</sup> E. W. Taylor, *Boston Med. and Surg. Jour.*, 1903, cxlviii, 634.

meatus, including the anterior part of the drum, a part of the cheek, lower lip and chin, the lower teeth, gums, and tongue as far back as the circumvallate papillæ, the floor of the mouth, the inner surface of the cheek, and salivary glands. The *motor portion* of the third division supplies the muscles of mastication, the masseters, temporals, both pterygoids, also the tensor tympani, the mylohyoid, and the anterior belly of the digastric. One of the most important anatomical relations of the fifth nerve is with the chorda tympani, through which the special sense of taste in the anterior two-thirds of the tongue is subserved.

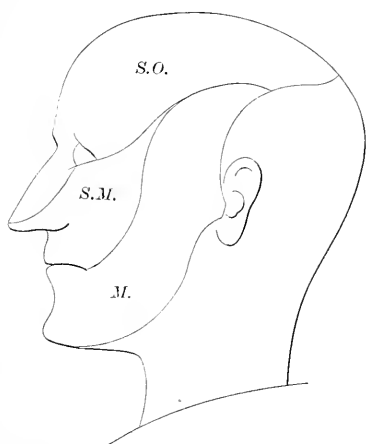
As Cushing<sup>1</sup> pointed out, a valuable means of determining the exact distribution and function of the trigeminal nerve in all its branches is to be derived from clinical study, particularly through the frequent operations on the Gasserian ganglion or more peripheral portions of the nerve. Cushing's studies are of the greatest value, and determined with

much accuracy the fields of anesthesia in the distribution of the nerve.

**Special Pathology.**—Owing to the long course and complicated ramifications of the fifth nerve, together with the fact that from origin to terminations it passes through several bony canals, it is natural that it should be damaged in various injuries or diseases of the brain and skull. It is, however, shown from Thomas<sup>2</sup> statistics that the nerve is infrequently involved in fracture of the skull. In a series of 69 cases he found it affected but once, whereas the facial nerve, either alone or in combination with other nerves, was involved in 44 cases. In disease of the pons, occasioned by hemorrhage, softening, tumor, multiple sclerosis, or abscess, the fifth nerve is naturally involved. Extensive affection of the

base of the brain is likewise often a cause of disturbance in the distribution of the fifth nerve. In the periphery a true primary neuritis of this nerve is exceedingly rare, and a neuritis secondary to disease in the neighborhood is also infrequent. Involvement of the nerve is naturally frequent in traumatism of various sorts, but it is rarely damaged in ordinary skull fracture. The pathological anatomy underlying the frequent neuralgia of the nerve is as yet wholly indefinite. Its association with arteriosclerosis, particularly in the neighborhood of the Gasserian ganglia, no doubt has some significance, but certainly in many cases is an insufficient and incomplete explanation of the pain. The studies of Spiller, Barker, Rusk, and Cushing have shown certain alterations in removed Gasserian ganglia, but not sufficiently characteristic to permit

FIG. 28



Cutaneous distribution of fifth nerve. S.O., supra-orbital division; S.M., superior maxillary division; M., mandibular division.

<sup>1</sup> *Jour. Am. Med. Assn.*, 1905, xliv, 773.

<sup>2</sup> *Ibid.*, 1908, li, 271.

of their definite association with neuralgia. The situation regarding the pathological anatomy of a somewhat frequent herpes occurring in the branches of this nerve is more definite. There seems little question that an involvement of the ganglion is the direct cause of the eruption in this as in other sensory nerves, and that the somewhat fanciful term posterior poliomyelitis is not wholly misplaced. Barker's and Cushing's experiments, with the Nissl method, showed that distinct groups of cells may be involved leading to localized herpes in the same sense that groups of muscles are involved in poliomyelitis of the motor type.

**Symptoms.**—By all means the most important affection of the fifth nerve from the practical standpoint is the so-called *tic douloureux*, or neuralgia in other forms. In the general symptomatology of the affections of the nerve primary symptoms of irritation, manifested by pain, should be recognized. A more serious involvement of the nerve leads to anesthesia in parts supplied, including certain mucous membranes. In such an anesthesia of one nerve, for example, a cup from which one is drinking feels as if broken. Food collects within the anesthetic cheek in spite of the fact that the motor power may be unimpaired. Taste is impaired over the anterior two-thirds of the tongue and somewhat at the back of the tongue, but does not remain wholly lost. The sensory loss to the facial muscles causes a peculiar awkwardness, due to an abolition, partial or complete, of the muscle sense. This disturbance not infrequently, particularly after neurectomies, may closely simulate a Bell's palsy. Cushing quotes such an instance in one of his cases of removal of the Gasserian ganglion. Such a condition is temporary and is easily determined by the persistence of normal electrical reaction. In considerable injury of the nerve the secretion of tears, of nasal mucus and of saliva is decreased, resulting in dryness with possible secondary trophic changes. Under these conditions the sense of smell may be lost finally through such changes in the Schneiderian membrane. The corneal, lachrymal, and palatal reflexes are lost, and the tongue lies on the paralyzed side. The teeth are anesthetic and tend to drop out. The condition of neuro-paralytic ophthalmia, after operation on the fifth nerve, has been regarded as a trophic change. This Cushing is inclined to doubt, since he has found that a partial neurectomy is more often a cause of this ophthalmia than a complete one, and that if the anesthetic eye be properly protected, such an ophthalmia does not occur. He attaches, therefore, particular importance to external sources of irritation.

**Taste.**—A wide difference of opinion exists regarding the course of the taste fibres and the part played by the fifth nerve in the transmission of gustatory sensations to the brain. There is general agreement that the glossopharyngeal nerve supplies the posterior portion of the tongue, and that the chorda tympani presumably subserves the same function for the anterior two-thirds. A difference of opinion arises as to the connection between the chorda tympani and the brain, whether it passes by way of the trigeminus or by the facial or glossopharyngeal route. The theory that the fifth nerve is the course over which the taste fibres pass has had a large following. According to this theory the course of the fibres is through the lingual, the chorda tympani, and the facial to

the geniculate ganglion, thence by the great superficial petrosal and sphenopalatine ganglion to the second division of the fifth nerve, and so to the Gasserian ganglion and the brain, or possibly through the small superficial petrosal and otic ganglion to the third division of the fifth nerve. Krause and Cushing arrive at somewhat different conclusions from their neurological studies of operative cases. This no doubt is due to a number of causes, among which, as Cushing points out, the failure to make adequate preliminary tests for taste, uncertainty regarding total extirpation of the ganglion, faulty methods of making the tests, or, finally, a failure to take into consideration the loss of common sensation in the part of the tongue under examination, are important. In thirteen cases which Cushing reported in 1903, he found that in only one was there a failure to reestablish taste perception, and this was found to be due to the fact that the chorda tympani had been divided at a previous operation and had not reunited. From this evidence it appears somewhat conclusive that the taste fibres do not, or do not exclusively, enter the skull along with the trigeminal nerve. The other alternative is that the fibres of the chorda tympani, after passing through the geniculate ganglion, reach the brain by way of the pars intermedia along with the facial nerve, constituting its sensory portion. The question is further complicated by the fact that lesions of the facial nerve involving the pars intermedia between the geniculate ganglion and the pons usually at least do not lead to affections of the sense of taste. The evidence, therefore, remains conflicting. The whole subject has been discussed by Davies.<sup>1</sup> The posterior third of the tongue and palate is presumably supplied in great measure through the glossopharyngeal nerve. Even from this part of the tongue, however, some of the fibres apparently pass to the brain by way of the fifth nerve, since disease of the Gasserian ganglion has caused impairment of taste, both in the anterior and posterior part of the tongue. The route of these fibres, therefore, has been presumed to be from the glossopharyngeal nerve to the fifth, through the tympanic plexus (Jacobson's nerve) to the small superficial petrosal, and so through the otic ganglion to the third division of the fifth. Cassirer reported a case of complete loss of taste from disease of the glossopharyngeal nerve. From this observation the possibility is suggested that the glossopharyngeal may be the special nerve of taste. It must, furthermore, be borne in mind that individual differences may exist and that the taste fibres may conceivably take different courses in different persons.

**Motor Portion.**—Paralysis or disturbance of the innervation of the motor portion of the nerve naturally occurs only in involvement of the third division. In extirpation of the Gasserian ganglion the motor division is necessarily destroyed, but without leading to serious consequences. Paralysis of the muscles of mastication takes place on the affected side—masseter, temporal, and pterygoids. Paralysis of the mylohyoid and anterior belly of the digastric do not lead to significant disturbance of function. Chewing is in general not seriously interfered with, owing to the preserved action of the corresponding muscles of the

<sup>1</sup> *Brain*, 1907, xxx, 219.

other side. The reaction of degeneration occurs in these muscles, but is rarely tested. Atrophy later supervenes. The lower jaw can be moved only toward the injured side, either on opening or on attempted lateral movement. There is also interference with wide opening of the mouth (digastric, mylohyoid). The paralysis which occurs in violent trigeminal pain is naturally merely apparent and due to pain inhibition. Paralysis from central lesion is rare, and the definite location of a cortical centre is as yet uncertain.

**Trismus.**—Tonic and clonic spasm of the jaw muscles, known as trismus, is rather a symptom than a specific affection. In the tonic variety the jaws are closely approximated. This occurs as a symptom of tetanus, in meningitis, seldom in tetany, as a temporary symptom in the epileptic seizure, also in certain diseases of the pons, and possibly as a result of irritation of cortical centres. It is also not very uncommon in hysteria, and may occur as a reflex from carious teeth or periostitis of the jaw. The clonic form of trismus manifests itself in rhythmic movements of the jaw, usually in a vertical, less often in a horizontal direction. It occurs in general convulsions, hysteria, epilepsy, and paralysis agitans. Extensive involvement of the central motor tracts may lead to a jaw clonus similar to that often obtained in the ankle. The prognosis of trismus is, as a rule, good, provided the underlying source of the affection can be alleviated or removed.

### FACIAL (SEVENTH) NERVE.

**Facial Paralysis.**—**Etiology.**—A paralysis of the seventh nerve is more frequent than that of any other nerve in the body. In the large proportion of cases it is unilateral, and the lesion producing it is usually in its peripheral distribution. Among the apparent causes of the ordinary type of peripheral facial paralysis exposure takes a prominent place. It has been estimated that 70 per cent, of all cases show this etiological factor. Due largely to the investigations of Reik<sup>1</sup> in this country, attention has been called to the relationship between non-suppurative otitis media and facial paralysis of the refrigeratory type. In Reik's view a middle-ear disturbance always precedes this variety of facial paralysis, causing the paralysis through direct inflammation of the nerve, or by pressure from exudate. Other suggestions have been made and theories of an infectious process, a neuritis, or a primary degeneration of the nerve within the Fallopian canal, all have advocates. In this connection a pathological report by André Thomas,<sup>2</sup> in which he was able to study three cases, is of interest. The cases were all of peripheral paralysis of the seventh nerve, two of them showing hemispasm. In each of these cases an examination of the nerve and of the nucleus was made. In the first case death resulted in eighteen days and the nerve showed a parenchymatous degeneration downward in the aqueduct

<sup>1</sup> *Johns Hopkins Hosp. Bull.*, 1902, xiii, 83; *Trans. Am. Otolog. Soc.*, 1904, viii, 365; *Internat. Clinics*, 1909, ii (19th Series), 221.

<sup>2</sup> *Rev. Neurolog.*, 1907, xv, 1273. See also A General Discussion and Bibliography on Facial Paralysis, A. Fuchs, *Arch. a. d. Neurolog. Inst. a. d. Wien. Univ.*, Band xvi, Part 2, 245.

of Fallopius from the first bend, with almost entire loss of the axones. Above the geniculate ganglion the axones were less altered. The cells

FIG. 29



Complete right peripheral facial paralysis. Following operation for sarcoma of the mastoid region, with unavoidable destruction of the seventh nerve. Impairment of taste, anterior portion of the tongue on the left. Face at rest, showing slight deformity. (Massachusetts Charitable Eye and Ear Infirmary and Massachusetts General Hospital.)

FIG. 30



Same as Fig. 29; attempt to close eyes.  
Right side of face immovable.

FIG. 31



Same as Fig. 29; attempt to show teeth.  
Paralysis of zygomatici and other muscles concerned in raising the lips.

of the facial nucleus were swollen and showed chromatolysis and eccentric nuclei, whereas the cells of the opposite nucleus were normal. In the



second case, following an otitis media, there was total left facial paralysis, with complete reaction of degeneration and occasional muscular spasm. Examination of this nerve showed chronic perineuritis below the first bend of the aqueduct of Fallopius. The nerve showed a neuroma of regeneration. The geniculate ganglion was somewhat atrophied, the facial nucleus normal. The value of this case is impaired by the pre-existing otitis. In the third case there was almost complete right facial paralysis with occasional spasm. Examination of the nerve showed signs of regeneration. There was swelling of the nerve at the first bend of the aqueduct of Fallopius. The nucleus was normal. In all of these cases the initial lesion in the aqueduct of Fallopius appeared at the first bend of the facial nerve.

The attempt has been made to hold a neuropathic tendency responsible for facial paralysis, but statistics do not support this. There are, however, instances in which cases have occurred in one family, and others in which recurrences have taken place. An emotional shock has been held responsible in occasional cases. Gowers reported the case of a woman who underwent a facial paralysis immediately after watching the dressing of a breast cancer, and Waterman has made somewhat similar observations. Constitutional diseases, such as gout, diabetes, leukemia, and syphilis, have all been held responsible. It has also occurred during the puerperal state and following diphtheria. It is rare as an accompaniment of a general polyneuritis, and if it occurs in this or in allied conditions, paralysis is apt to be double-sided, as the writer has observed in several cases. A facial paralysis may also, no doubt through a nuclear lesion, occasionally occur in poliomyelitis. One such case was observed in a child, in whom the only other involvement was a slight paralysis of the anterior muscles of one leg. We are on much more definite etiological ground in considering the relation of trauma. Owing to its long course through the Fallopiian canal, and its close proximity to a cavity which often suppurates, paralysis of a facial nerve from carious and suppurative ear disease is extremely common. Basal fractures likewise show a facial paralysis in a large proportion.

Naturally disease of the *base* of the brain, meningitis, new-growths, and affections of the medulla and pons, may readily lead to involvement of one or both facial nerves. The occasional palsy of a branch of the facial nerve supplying the orbicularis oris in cases of bulbar palsy, presumably through the degeneration of fibres passing out with the twelfth nerve, is of interest. The observation which Cushing has made, that certain of the cases of supposed paralysis of the seventh nerve, following extirpation of the Gasserian ganglion, are due rather to loss of muscle sense in the fifth nerve than to an actual paralysis of the seventh, is likewise worthy of mention. In an analysis of 335 cases of facial paralysis, exclusive of those due to ear disease or trauma, G. A. Waterman<sup>1</sup> arrives at the following conclusions relative to the etiology and course. He finds that sex plays no significant part in the incidence; that hereditary influence is doubtful; that exposure to cold is the most definite etiological

<sup>1</sup> *Jour. Nerv. and Ment. Dis.*, 1909, xxxvi, 65.

factor as yet determined; that the affection is no more frequent in winter than in summer; that it may occur at any period of life, but is much more frequent before forty and during the third and fourth decades. Thirty-six per cent. of all these cases occurred between twenty and thirty. The disease is more severe, but less frequent in later life. Preliminary pain is not prognostic, and secondary contractures are not affected by galvanic treatment, as has at times been maintained.

**Onset and Symptoms.**—The onset of an ordinary peripheral facial paralysis is usually rapid and at times sudden. Naturally the signs are produced more slowly if the paralysis is due to a gradually increasing disease of the middle ear, or to a progressive tumor growth. Pain is a not unusual prodromal symptom, located in the ear, behind the ear, in the mastoid region, or in the neck, but is of small diagnostic or prognostic significance, according to Waterman's statistics. The explanation of this preliminary pain is not easy. It may be due either to coincident involvement of sensory branches or possibly to a neuritis which may be regarded as an accompaniment or cause of facial paralysis. At times the facial paralysis is ushered in with fever and signs of infection, especially in children. In the absence of a slowly progressive cause, a gradual onset of the paralysis is extremely unusual. The signs of facial paralysis are naturally conspicuous. The most striking is a loss of movement, often complete, of the side of the face affected, more marked in elderly persons than in young, no doubt due to the presence of wrinkles in advancing years, which when obliterated by paralysis increase the contrast between the normal and affected side. The eyelid remains open, to a certain extent even in sleep, owing to the paralysis of the orbicularis oculi. In an attempt to close the eye the globe is rolled upward, showing the sclerotic and often leading to the false idea on the part of the patient that the eye is completely closed. The partial closure of the eye often observed in the presence of very complete paralysis of other branches of the nerve is due to relaxation of the levator rather than to any active contraction of the orbicularis. In giving expression to the emotions, as in smiling, the mouth is drawn toward the unaffected side through the unopposed action of the zygomatici. The tongue appears to be protruded to one side, due to the fact that the position of the mouth is altered by the paralysis. The tongue is in reality not affected. The lips cannot be properly apposed, so that whistling is not possible. Drinking is likewise interfered with, and speech may suffer from the impossibility of properly making use of the labial sounds. From the paralysis of the buccinator, food often remains between the teeth and in the cheek until mechanically removed. The muscles of the external ear are paralyzed, as well as the platysma, so that the lower lip cannot be drawn down. The palate is not affected.

In considering the point at which the nerve is involved, a somewhat artificial distinction may be made regarding five positions (Fig. 32):

1. If the lesion lies below the exit of the nerve from the stylomastoid foramen, taste will not be affected, inasmuch as the chorda tympani joins the nerve above this point.

2. If the lesion be between the stylomastoid foramen and the geniculate

ganglion within the Fallopian canal, taste in the anterior two-thirds of the tongue is affected through coincident involvement of the chorda tympani, which accompanies the facial nerve between these points. In addition there is also occasional slight loss of general sensibility, with possible subjective taste sensations and changes in the secretion of saliva. The tongue may be furred in consequence of the altered salivary secretions. If the nerve to the stapedius muscle given off from the trunk of the facial nerve, between the geniculate ganglion and its exit from the

FIG. 32

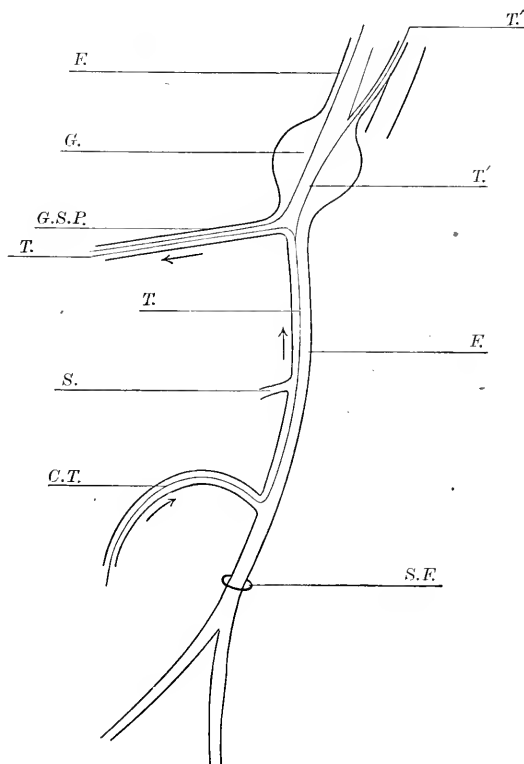


Diagram of facial nerve. *F.*, facial root; *G.*, geniculate ganglion; *G.S.P.*, great superficial petrosal nerve; *S.*, nerve to the stapedius muscle; *C.T.*, chorda tympani nerve; *S.F.*, stylomastoid foramen; *T.T.*, taste fibres; *T.T.T.*, possible course of taste fibres through the pars intermedia. The arrows represent the probable course of these fibres. Lesions of the nerve from the geniculate ganglion to the point at which the chorda tympani is given off lead to disorder of taste, with probable impairment of hearing.

skull, be involved, hypersensitiveness to musical notes of low pitch has been observed.

3. If the nerve is affected between the geniculate ganglion and the pons, there is in general the same symptomatology as in the ordinary peripheral type. There is no involvement of taste, which has been used as a somewhat forcible argument against the theory that the pars intermedia carries the taste fibres. Deafness is often associated with lesion of the facial nerve at this point from the fact that the auditory nerve

accompanies it. A deficiency of tear secretion on the affected side has been described in many cases, and possibly an involvement of the stapedius.

4. If the lesion is in the pons itself, taste and hearing are unaffected, and there are also almost invariably lesions of other nerves, notably of the sixth, accompanying the facial paralysis.

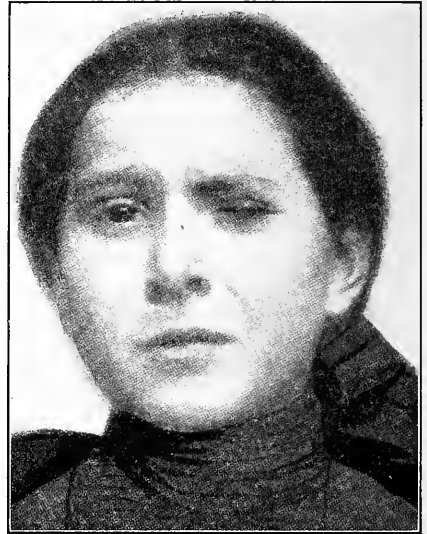
5. If the central course of the facial nerve be involved, the symptomatology differs from all those previously considered, inasmuch as no changes in electrical reactions occur and the upper branch of the nerve is very much less involved than the two lower.

FIG. 33



Left facial paralysis of long standing. Showing inability to close eye and laxity of muscles, especially evident about the mouth. (Massachusetts General Hospital.)

FIG. 34



Spasm of left orbicularis oculi. Photograph shows an attempt to open the eye, which tends merely to increase the spasm; girl, aged twelve years. (Putnam and Waterman. *Studies in Neurological Diagnosis*. Massachusetts General Hospital.)

The facial nerve is a peculiarly easy one to examine with the electrical current, and the information gained from this source is of the greatest possible value in arriving at a diagnosis. The presence or absence of electrical changes may be the determining factor as between a lesion peripheral to the pons or above the pontine nuclei in the central course of the nerve. Altered electrical reactions naturally occur only in lesions of the peripheral neurones. In slight degrees of paralysis the electrical reactions may be unaffected. In severe cases a complete reaction of degeneration may be established in two weeks. Between these two extremes forms of partial reactive degeneration may occur, and upon the information thus given depends very largely an estimate of the duration of the attack.

**Diagnosis.**—It will be seen that the diagnosis of the approximate position of the lesion is usually easy. It is especially important often to distinguish between a central disturbance in the nerve and one due to a peripheral cause. The essential points in making this differential diagnosis are as follows: In the central lesion there is no change in electrical reactions; the upper branches of the facial nerve are slightly involved, either due to the fact that there are special cortical centres for different portions of the nerve, or, more probably, because muscles which ordinarily act together are presumably innervated from both sides of the brain. The facial paralysis is usually accompanied by a homolateral hemiplegia. A crossed or alternating hemiplegia indicates a lesion in the pons below the crossing of the central facial fibres and above the crossing of the pyramid. The reflex conditions of the nerve are theoretically increased, but for practical purposes may be regarded as unchanged. In contrast to this series of conditions, lesions of the peripheral nerve, irrespective of special location, are, in general, as follows: Electrical alterations are always present, except in the mildest cases. All branches of the nerve are equally involved, and reflex activity is diminished, with ultimate muscular atrophy, if recovery does not take place. A modification of the foregoing general statement must naturally be made in those instances where through trauma a special branch of the nerve has suffered.

**Course and Prognosis.**—The outcome, as well as the course, varies within wide limits. Slight cases may last not more than one or two weeks, and give very little physical evidence of their presence, either by inspection or on physical examination. Other cases may continue for weeks, months, or even become permanent. As already suggested, the electrical conditions are the most important determining factor in relation to prognosis. A complete reaction of degeneration is by no means inconsistent with final recovery, but a reaction of degeneration persisting for months, and finally giving place to a quantitative diminution of electrical excitability, must always be regarded as a grave prognostic sign, indicative of permanent changes in the nerve, usually no doubt due to a greater or less degree of destruction of the pontine nucleus. A somewhat artificial distinction may be made between a mild, more severe, and a severe form of paralysis, depending upon whether the electrical reactions are normal or very slightly affected, with recovery in two or three weeks, or, in the second case, whether a partial reaction of degeneration is present, with recovery in from four to eight weeks, and finally, in the severe form, in which the reaction of degeneration is complete, with a duration of paralysis from three to six months. Naturally there are certain cases which do not conform to this classification, as, for example, those in which the paralysis lasts many months, or those in which the electrical reactions do not correspond with the apparent degree of paralytic defect.

In the ordinary case, after a varying period of complete reaction of degeneration, the electrical alterations change for the better in that the reaction of degeneration is no longer complete. The cathodal contraction increases over the anodal, and the faradic irritability of nerve and muscle is gradually reestablished. Along with this improvement in electrical

reactions voluntary control of the muscles is gradually regained until the face is restored to an essentially normal condition.

*Secondary Contractures.*—In giving a prognosis in peripheral facial paralysis the possibility of secondary contractures should always be borne in mind. Even in the cases of moderate severity a certain amount of defect may persist in the movements of the face on the affected side and in the tendency of the muscles to overact. Such secondary contractures usually come on from four to six months after the onset, and not infrequently lead to a false conclusion as to the affected side. Inasmuch as the tendency is toward overaction, the affected side of the face shows a somewhat general contracture, the aperture between the lids is diminished, and on smiling or other emotional expression the affected side of the face is much more actively used than the unaffected. A further annoyance is the onset of spontaneous twitching of the facial muscles, often with isolated spasmodic contractions, especially of the zygomatici. This condition often persists for years, and, in fact, is not infrequently permanent, and may well prove to be a very much more disturbing condition than the original paralysis. Operation for its relief has been attempted with some degree of success. The cause of these secondary contractures has been a subject of much discussion, and is not yet definitely determined. A favorite theory has been that in any serious facial paralysis the nucleus of the pons is of necessity involved, the cells suffer and degenerate beyond the point of restitution, thereby interfering permanently with adequate innervation. Nerve impulses are conceived as spreading from the damaged nucleus in an irregular and somewhat uncontrolled manner, so that an excessive degree of a normal phenomenon is produced in the contracted parts.

A lessened resistance between the cells of the nuclei has also been suggested. A plausible theory also is that which regards the contractures as due to permanent alterations in the muscles of such a character that the fibrillar substance is imperfectly reproduced, whereas the sarcoplasm maintains the muscle in a state of exaggerated tonus.

In other than the ordinary form of facial paralysis, which has been especially considered, prognosis depends essentially upon the underlying cause, and is usually a relatively insignificant matter in view of the more serious condition of which it is a sign. In tumor, for example, and similar conditions, except for its localizing and, therefore, diagnostic value, a lesion of the seventh nerve naturally depends for its outcome upon the course of the growth which produced it. Certain cases have been described as recurrent, not unlike periodic oculomotor palsy. Several cases have also been described as occurring in one family, suggesting some hereditary predisposition. Both of these circumstances are too infrequent to alter the general statement that peripheral facial palsy of the so-called rheumatic type is a benign affection of good prognosis.

*Treatment.*—This consists in various measures of assisting nature in what may be regarded as a natural process of cure. Not too much, therefore, should be expected of any definite course of medical treatment, although as an adjuvant such treatment is unquestionably often useful and at times demanded. In view of the fact that a neuritis within the

Fallopian canal may be regarded as a pathological basis of the affection, fomentations and blisters may be applied in the mastoid region to serve as counter-irritants. If Reik's view be accepted that an otitis media always precedes a paralysis of the so-called refrigeratory type, an early paracentesis of the drum membrane should lead to rapid cure, and this has been the result in cases reported by him. The bowels should be kept free, the salicylates may at times be administered with advantage, and, if there is reason to regard syphilis as an etiological factor, the iodides, salvarsan, and mercury are indicated. The chief reliance in the treatment of this paralysis has been the use of electricity, and in spite of its recognized limitations, this form of treatment remains in popular favor. It is doubtful whether the electrical current has any specific effect upon the nerve, but it cannot be questioned that the stimulation of muscles incapable of movement through an act of the will is a reasonable and logical procedure. Oppenheim believes that in early, and often in older, cases striking results may be obtained by the use of electricity. In treatment by this means, presuming that a reaction of degeneration is present, the galvanic current should be employed, using over the affected muscles the pole which produces the more marked contraction. Not more than 4 to 5 milliampères should be used, and the muscles should be stroked by an electrode for a period not exceeding ten minutes. As the faradic irritability returns, this form of electricity may be substituted for the constant current, but it is well to discontinue any form of electrical treatment when the tendency to contracture and spasm of the muscles becomes apparent. There is no adequate evidence to show that the application of electricity plays any part in the production of secondary contracture, but the treatment becomes more and more superfluous as voluntary control over the muscles increases, and may well be discontinued before such voluntary control becomes complete.

By surgical intervention intractable cases of facial paralysis may be very greatly benefited, if not completely cured. The first method, suggested by Ballance in 1895, was the anastomosis of the facial with the spinal accessory. In this operation a part or a whole of the spinal accessory was sutured into the divided facial, or an implantation of the facial nerve made into the split spinal accessory, the object being to facilitate the growth downward of spinal accessory fibres in the path of the functionless facial. Following Ballance, cases of this operation were reported by Faure, Kennedy, Cushing,<sup>1</sup> and Hackenbruch. The immediate result was a more or less permanent paralysis of the trapezius and sternomastoid muscles. The facial condition improved, but any sudden voluntary movement of the face was accompanied by a distressing involuntary movement of the shoulder and *vice versa*, which proved to be a decided disadvantage. To obviate this the hypoglossal nerve was selected for the anastomosis. Inasmuch as the hypoglossal nerve is closely associated physiologically with the facial, and since the cortical localization of the two nerves is near together, it seemed more rational to use the hypoglossal than the spinal accessory and a considerable number of such operations have been done.

<sup>1</sup> *Annals of Surgery*, 1903, xxxvii, 641, with historical and bibliographical notes.

The importance of operating as soon as possible after injury of the facial nerve can hardly be too strongly insisted upon, inasmuch as the progressive disintegration of muscle may easily go on to a point where recovery is no longer possible, however completely the nerve may regenerate. In carrying out this operation it is desirable not to divide the hypoglossal completely, owing to the consequent difficulty of mastication, deglutition, and phonation. Indication for the operation is the assurance that the facial nerve is divided or so completely injured that spontaneous recovery is impossible. Its field, therefore, lies essentially in those cases in which the nerve has been divided through trauma, particularly resulting from accidental injuries in operating on the ear, and from destructive suppurative processes within the middle ear.

**Facial Spasm and Tic Convulsif.**—Irrespective of the postparalytic contracture and spasm, the facial nerve is peculiarly prone to spasmodic affections, due, no doubt in part, to its complex reflex relations. There is, in the first place, an exceedingly close relation between the mental condition and the muscles of expression, and in the second place the reflexes which subserve this relation are naturally highly complex. A localized irritation of the nerve may lead to spasm, which later, when the cause has been removed, may persist as a fixed habit. The direct irritation of the nerve stem, as by a tumor, or the occasional irritation from a localized brain lesion, may produce more or less persistent facial spasm. In most of the cases no definite anatomical basis is demonstrable. Brissaud and especially Meige have distinguished between what they regard as true spasm and one of psychogenic origin. Of the latter, are those aroused emotionally, which later have become habitual. Such are various reflex spasms, the so-called *tic convulsif*. As distinguished from this type the true facial spasm is not dependent upon a mental state, but is due rather to material changes in the reflex arc either in the sensory trigeminus, facial nucleus, or the facial stem. The first type bears close relation to the psychic life, and may be externally influenced through diversion of the attention, and by similar means. The second type of spasm usually occurs in the whole distribution or in definite portions of the nerve, and is usually one-sided, and presumably is not influenced by suggestive measures. This type of spasm is that artificially induced by an electric current. A true cramp of the facial nerve may likewise occur, either tonic in character or alternating.

Patrick<sup>1</sup> made clear the fundamental difference between true facial spasm and tic. He states the distinction as follows: "Spasm may be said to be an anatomical, tic a physiological, disorder. A good picture of facial spasm may be obtained by faradization of the facial nerve. Tic is always the replica of a perfectly natural (though may be unusual) and physiological movement. Looking at the distortion caused by faradization of the nerve no one would mistake it for voluntary contraction of the facial muscles. On the other hand, having seen only one individual contraction of facial tic, no one could say that the movement was not a perfectly normal and natural one—possibly unusual, possibly overdone,

<sup>1</sup> *Jour. Nerv. and Ment. Dis.*, 1909, xxxvi, 1.



but the legitimate result of a casual cause. Voluntary imitation of facial spasm is practically impossible. The patient himself can always repeat or imitate his tic movements, and another person can nearly always do so." In so-called blepharospasm, one of the commonest of the spasmodic affections, the subjective disturbance may be extremely slight or wholly lacking, especially in its milder forms.

In so-called tic convulsif there are often signs of other nerve lesions and even of a definite underlying hysteria. The course of these affections is chronic, the prognosis doubtful but never hopeless.

**Treatment.**—This consists in the removal of any reflex cause. Drugs are, on the whole, useless, except as palliatives. Nerve stretching is possible, but should only be resorted to in very intractable cases. Injections of alcohol into the facial nerve near its exit from the skull have been used in intractable cases of facial spasm. Patrick reports three such cases, with excellent results in two and with a failure in one due to faulty technique. The treatment consists in the injection of a 40 to 75 per cent. alcohol solution into the trunk of the facial nerve. If the injection is successful, an immediate paralysis of the nerve results, naturally with a cessation of the spasm. Recovery takes place from the paralysis, and there is reason to believe that the spasm will not return even after a complete restoration of the normal function of the nerve.

Occasionally a spasm of the platysma may develop. Such a case has recently come under personal observation in which the patient, a man, for two or three years has been annoyed by a distinct spasm of the right platysma when talking, without apparent involvement of other muscles supplied by the facial nerve.

**Herpetic Inflammations of the Geniculate Ganglia.**—Under this general heading J. Ramsay Hunt<sup>1</sup> has brought together a group of cases which he regards as constituting a hitherto undescribed clinical entity. He regards the facial as a mixed nerve, the nerve of Wrisberg being its sensory portion, with the geniculate as its ganglion of origin. It follows that herpes may occur in its sensory distribution in common with other ganglia of this type, as, for example, the Gasserian ganglion of the fifth nerve or the various spinal ganglia. Hunt points out that the zoster zone for the geniculate ganglion lies in the interior of the auricle and in the external auditory canal. On the clinical side the affection manifests itself in the characteristic way, with slight febrile prodromata, neuralgic pains in the neighborhood of the ear, followed on the third or fourth day by the typical herpetic vesicles, very rarely, according to Hunt, localized on the tympanic membrane. This area retains its sensation after extirpation of the Gasserian ganglion and the second and third cervical ganglia, from which it is assumed that it represents the cutaneous distribution of fibres of the geniculate ganglion.

Herpes zoster of the auricle has long been recognized, but had heretofore been regarded as belonging to the trigeminal area, and hence due to involvement of the Gasserian ganglion. Hunt's position seems well taken that this localized manifestation of herpes may be more properly

<sup>1</sup> *Jour. Nerv. and Ment. Dis.*, 1907, xxxiv, 73; *Am. Jour. Med. Sc.*, 1908, cxxxvi, 226.

explained by an involvement of the nerve of Wrisberg and its ganglia. The cases of herpes in and about the ear are uncommon. Nevertheless, in a very large collection of cases from special hospitals a certain small number have been reported for many years. Hunt described three clinical types: (1) Uncomplicated auricular herpes; (2) auricular herpes with facial palsy; and (3) auricular herpes with facial palsy and auditory symptoms. The latter group of cases is perhaps the most obscure and difficult of classification. On the basis of 60 personal observations and a somewhat limited pathological investigation, Hunt feels justified in establishing a characteristic syndrome divided into the three foregoing clinical groups. He also makes the statement that neuralgic affections of the geniculate ganglion and its divisions productive of an otalgia may properly be regarded as due to disorder in the sensory mechanism of the facial nerve, giving rise to earache of non-inflammatory origin. Anatomically, the geniculate ganglion stands in relation to the interior of the auricle and external auditory canal, to the tympanic plexus, to the second and third divisions of the fifth nerve, to the glossopharyngeal, and to the vagus. It is evident, therefore, that ear pain may be due to a variety of reflex causes. Hunt is convinced that, although many of the neuralgic affections of the ear are no doubt dependent upon involvement of the trigeminal and occipitocervical nerves, there exists also an otalgia due entirely to involvement of the sensory branch of the facial nerve.

Orbison<sup>1</sup> takes issue with some of Hunt's conclusions and makes a still further division of zoster zones on the basis of a study of herpes of the tympanic membrane. He believes that herpes of this area is due not, as tacitly assumed by Hunt, to involvement of the zoster zone of the seventh nerve, but rather to zoster conditioned by disease of the petrosal ganglion of the glossopharyngeal.

### AUDITORY (EIGHTH) NERVE.

**The Cochlear Nerve.**—The function of hearing is almost undoubtedly subserved wholly by the cochlear division of the auditory nerve. Hearing may be affected through lesions anywhere in the course of the nerve, from the cortex through the various sensory nuclei to the labyrinth, but such disturbance is far more frequent in actual disease of the internal ear and its structures and in affections of the terminal branches of the nerve than in more centrally localized lesions.

1. The cortical centre for hearing is presumably localized in the superior convolution of the temporal lobe, and unquestionably stands in close relation with the cortical area for the perception of language. There is, however, still disagreement regarding its exact position. The reason that word deafness does not lead to sound deafness is that the auditory nerves naturally have relations with both hemispheres, whereas the more highly specialized function of the appreciation of language is confined wholly to one hemisphere. The observation has been made that when both superior temporal convolutions are diseased, sound deafness has

<sup>1</sup> *Jour. Nerv. and Ment. Dis.*, 1908, xxxv, 500.

also resulted. Inasmuch as this latter lesion is extremely unusual, it follows that deafness rarely results from cortical lesions.

2. Lesions of the auditory nerve from the cortex to its subsidiary nuclei may also lead, under certain circumstances, to deafness which, on the practical side, is indistinguishable from deafness due to labyrinthine disease. Lesions of the corpora quadrigemina and geniculate bodies, tumors of the substance of the pons, also certain tabetic conditions and multiple sclerosis, may lead to deafness in one or both ears through involvement of the auditory mechanism in its course through the brain stem. More practically important are lesions in the peripheral course of the nerve. Inflammatory processes, notably meningitis, are likely to involve the nerve with others at its exit from the pons, often spreading along the nerve to the ear itself. A primary neuritis of this nerve and subsequent atrophy, also the so-called rheumatic paralysis similar to that frequently observed in the facial nerve, are exceedingly rare conditions, if, in fact, they exist. A condition of importance, to which special attention has been drawn, is that of tumor growing in the cerebellopontile angle. Such tumors are not infrequently neurofibromatous in character, and may under certain circumstances be removed. They are most frequently localized on the eighth nerve, but may also involve the trigeminus and vagus. Tumors lying in this region naturally give rise to auditory symptoms, which may aid in determining the location. Aneurism is at times a cause of deafness through pressure on the nerve.

3. In the great majority of cases the lesion inducing the deafness or other disturbances in the function of the auditory nerve lies in the internal ear, either as a primary process or resulting from the extension of disease from the middle ear. Causes of such disturbances are hemorrhage, inflammations, sclerotic processes often on the basis of acute infectious disease, particularly mumps or syphilis, certain constitutional diseases, as, for example, nephritis, diabetes, and pernicious anemia. A very common cause of labyrinthine disease is epidemic cerebrospinal meningitis, which not infrequently results in permanent deafness. Hysteria not infrequently shows a definite loss of hearing.

**Symptoms.**—These may be classed in two groups—symptoms of irritation and symptoms of defect. Irritative symptoms or hyperesthetic conditions are unusual, but have been observed in certain abnormal functional states of the nervous system, notably in hysteria. In this condition sounds may be distinctly appreciated which are not ordinarily heard. Such hyperacuteness of hearing may also be observed at the onset of certain acute and general diseases, and undoubtedly in these instances may be attributed to a general hyperexcitability of the nervous system. In certain disorders of the conducting sound mechanism, as, for example, in paralysis of the stapedius muscle often occurring in conjunction with a general facial paralysis, low notes may be heard with unusual distinctness, due presumably to the relaxation of the stapedius muscle. The condition of so-called *dysacusis*, in which sounds ordinarily innocuous cause distinct discomfort, should also be mentioned. Under these conditions pain, as, for example, headache, is apparently

actually increased by noise. Undoubtedly this affection, which is not an uncommon one in healthy persons, is due to a temporary general lowering of the nervous tone or to disturbance in the middle ear.

**Treatment.**—This must depend largely upon the underlying condition. If, for example, the stapedius muscle is involved conjointly with the facial nerve, the difficulty, which in any event is a trifling one, will pass off with the correction of the facial paralysis. The general nervous conditions which underlie the abnormal auditory phenomena must be treated. The bromides are undoubtedly useful, but much more important is a consideration of the general nervous and physical health.

**Tinnitus Aurium.**—Under this general heading are included various subjective sound sensations referred to the ear, the character of which may be roaring, hissing, ringing, ticking, or whistling. This affection in its varied forms is extremely common and often most distressing. Its causes are manifold. In consideration of the normal sensitiveness of the auditory mechanism, it is, in fact, remarkable that subjective sounds are not constantly being heard. One reason for this undoubtedly is that we come to ignore many sounds, which, if our attention were directed to them, would become exceedingly annoying. To city dwellers at any rate noise is practically constant, which is, for the most part, ignored. More in detail, some of the more important causes of tinnitus are:

Any morbid process in the ear may cause subjective sounds, but especially disease of the labyrinth, which is accompanied by more or less nerve deafness. Among these are alterations in circulation due to anemia, abnormal pulsation in the carotid artery, intracranial aneurism, or possibly vasomotor palsy of the labyrinthine vessels. Osler speaks of a bruit in the ear, with systolic intensification, which was supposed, for a time, to be due to an aneurism. Cerumen in the external meatus and various affections of the middle ear may be operative in producing tinnitus. The actual involvement of the nerve endings may also be responsible, in which case irritative processes or clonic spasm of the internal ear muscles may be operative. Violent stimulation of the auditory nerve by loud and particularly by continuous sounds may produce a persistent tinnitus. Workmen in boiler factories, foundaries, and similar places of employment are not uncommonly so affected. Finally, in various general nervous conditions—hysteria, neurasthenic states, in the aura of epilepsy, or in migraine—subjective sounds may be a prominent feature. A further elaboration of these simple forms of tinnitus leads often to very elaborate subjective sounds, which must then be assigned to the interposition of higher centres and probably are attributable to the action of the cerebral cortex itself. Such are the auditory hallucinations not infrequently observed in the insane. In all such conditions the relation of the mental attitude to the physical condition in the ear is naturally of very great interest and importance in modifying and keeping alive the condition. It is certainly true in this as in other neuroses that a definite cause in the ear leading to simple tinnitus may well produce a habit with which ultimately the original cause has nothing to do. If the tinnitus is double-sided the prognosis is much less favorable than in the unilateral affection.

**Diagnosis.**—In this very little information is to be gained from the character of the sounds. Although it is true that sounds synchronous with the pulse are probably directly or indirectly due to arterial pulsation, nevertheless such sounds may also be intermittent. It may, in general, be said that sounds of arterial origin are more or less continuous, but here again it is necessary to be on one's guard, since, for example, cerumen has been responsible for a constant tinnitus. The apparent location of the sound is also most varied. It may be referred to the ears, be bilateral or unilateral, or be referred to the head either at the occiput or vertex. The relation to deafness is also extremely vague, since at times the tinnitus may decrease with increasing deafness, or may persist unchanged.

It is evident that a definite diagnosis is often extremely difficult. If the sounds are elaborate it is usually safe to assume that they are of central origin, or at least that the cortex takes part in the general condition. The further question of the exact relation of the labyrinth to various subjective sounds must naturally be determined by expert otological examination. It is, however, well to note whether continuous sounds are increased or diminished by the recumbent posture. If the tinnitus is the result of venous hyperemia, it is ordinarily increased by lying down, but if due to simple anemia, it is relieved thereby. It should also be remembered that certain drugs, notably quinine and the salicylates, may produce a tinnitus, and that persons with middle-ear or labyrinthine disease are particularly susceptible. A pulsating tinnitus due to arterial congestion, may often be relieved by pressure over the carotid artery, and this may serve as a diagnostic hint.

**Treatment.**—This is usually unavailing unless its cause may be discovered, which is not infrequently impossible, and when possible the condition is often such that there is no available relief. The bromides and counter-irritation behind the ear may at times serve a useful purpose. If syphilis is suspected as a cause a vigorous treatment directed against that disease should be instituted. Tinnitus due to so-called functional disorders of the nervous system may be helped by the use of galvanism, using the anode as the active pole, and also by various suggestive measures. Recently Frazier<sup>1</sup> has reported a case of successful intracranial division of the auditory nerve for tinnitus, as suggested by Mills in 1908 for vertigo. This somewhat radical procedure should find favor in selected cases but only in the hands of specially skilled operators.

**Nerve Deafness.**—This term is applied to the inability to hear, conditioned by an affection of the cochlear nerve with accompanying labyrinthine involvement. The details of this subject lead directly into the field of the otologist, but certain tests must be made by the practitioner which will throw light on the general character and location of the disturbance leading to the deafness. For this purpose a watch with a fairly distinct tick, or, better, a middle-range tuning fork, is essential. In making such an examination it is first necessary to determine whether the external auditory meatus is clear of wax and the drum membrane intact. Having determined these points, one ear should be closed, the

<sup>1</sup> *Jour. Am. Med. Assn.*, 1913, lxi, 327.

patient directed to close his eyes, and the watch brought from a distance toward the ear, the exact point being noted at which the tick is first distinctly heard. It should be remembered that the tick is not heard at so great a distance under these conditions as when the watch is gradually removed from the ear. The results obtained should be compared with the other ear and with a normal control. Considerable variation will be observed, but relatively speaking the test is sufficient to demonstrate the presence or absence of conduction by air. Should such an examination be negative, it indicates simply that the external ear, the internal ear, and the sound-appreciating apparatus are intact; in other words, it forms the most general relative test of hearing. Under certain conditions the conduction through the air and through the internal ear to the cochlea may be interfered with by disease in such a way that sounds no longer penetrate by way of the air. It is, therefore, necessary to examine bone conduction. Under normal conditions a sounding tuning fork placed either over the mastoid or other part of the skull in contact with the skin will be heard in both ears. Inasmuch, however, as the conduction through the bone to the internal ear is less direct than through the air, it happens that the tuning fork, when no longer heard over the mastoid, if placed before the ear will again be heard. This is the usual course of events under normal conditions and in nerve deafness. If, on the contrary, the sound-conducting apparatus is diseased through a plugging of the external meatus or through disease of the middle ear the sound will naturally not be heard aërially after it has ceased to be heard through the bone. If, therefore, there is an affection of the auditory nerve leading to so-called nerve deafness, a tuning fork is not heard through the bone, inasmuch as the receptive mechanism of sound in the cochlea is diseased. Under these conditions it may or may not be heard aërially. This is known as Rinné's test. This may be summarized as follows: In labyrinthine disease both air and bone conduction hearing are diminished and bone conduction is usually lost; whereas, in middle-ear disease air conduction is diminished and bone conduction is increased.

As a further associated test of that devised by Rinné, the Weber test is ordinarily employed. Under normal conditions, if a tuning fork be placed on the forehead in the middle line and one meatus closed, the sound is best heard in the closed ear, since the aërial conduction is prevented on that side, whereas bone conduction is preserved and intensified through the conversion of the ear into a closed chamber. If, therefore, the sound-conducting mechanism is affected, the tuning fork is best heard on the affected side. This is known as a positive Weber test. If, on the other hand, nerve deafness exists from involvement of the labyrinth or the auditory nerve, the fork when applied to the forehead can no longer be heard on the affected side, inasmuch as the percipient mechanism is at fault. This is known as a negative Weber test. The Weber test, in general, is much less dependable than the Rinné test. These two tests are supplementary, and through their use a more definite knowledge may be obtained of the location of disease either in the sound-conducting mechanism, external auditory canal and middle ear, or in the sound-perception apparatus—labyrinth and cochlea. It is an interesting fact

that in nerve deafness hearing is usually worse in the midst of noise, whereas in deafness from chronic disease of the middle ear hearing is improved by external sounds.

**Vestibular Nerve.**—Disease of this branch of the auditory nerve, so far as it is separable from affections of the cochlear nerve, leads essentially to disturbances of coördination, of which vertigo, including Ménière's complex, is the most important. Owing to the relations which the vestibular branch bears to other cranial nerves and to the spinal cord various incoördinations of the muscles of the head, neck, and eyes may result from a lesion of the nerve. Nystagmus, for example, has been observed as one of these symptoms. Tinnitus, due to disturbance in the special sense of hearing, is often difficult to separate clinically from vertigo, of which it is often a symptom. The two conditions are very frequently associated, but it is nevertheless best to consider vertigo as a distinct disorder of equilibration in connection with the vestibular nerve.

**Vertigo.**—This may be defined as a sensation of giddiness and uncertainty resulting from a defect of equilibration, which is a complex act of muscular innervation, regulated ultimately by the cerebral cortex through the vestibular nerve and its connections in the ear and in the central nervous system. Experimental and other evidence goes strongly to show that the cerebellum is important in this function and that fibres of the vestibular nerve passing from the semicircular canals to the cerebellum are of great significance in the preservation of equilibrium. It is also clear that the semicircular canals, standing as they do in close relation to the vestibular nerve, are to be regarded as an essential part of this complex reflex. Finally, the cerebral connections, directly and indirectly, of the vestibular nerve bring about the cerebral control, without which preservation of equilibrium is impossible. It is perhaps too much to assume that the vestibular nerve is invariably concerned directly in the production of the symptom of vertigo, but that it indirectly takes part in this condition through its various reflex connections is, in our present state of knowledge, hardly to be questioned. The relation of the vestibular nerve to the ocular nerves and muscles is likewise of great importance, and accounts for the vertigo frequently observed in connection with defects in ocular innervation. The production of vertigo through affections of the larynx and various portions of the gastro-intestinal tract is no doubt anatomically explainable through connections with the vagus group of nerves.

Vertigo is described as *subjective* when the movement seems to be in the patient himself, and *objective* when external objects appear to be in motion. The usual result of a feeling of vertigo is the phenomenon of staggering. Inasmuch as vertigo occurs under many conditions, it is often a matter of extreme difficulty to arrive at an estimate of its significance in any given case. On the one hand, vertigo may be one of the symptoms of organic changes in the ear or central nervous system of gravest character, or, on the other hand, may indicate a nervous disturbance of the slightest sort. As Oppenheim pointed out, a distinct sensation of vertigo may be brought about in perfectly normal persons by closing the eyes, standing on one foot, and directing the attention to the general

uncertainty of this position. This fact, however, should not lead us to minimize its possible importance, particularly if associated with other symptoms of more or less grave character. In tumor of the brain, and particularly of the cerebellum, vertigo is a diagnostic point of much significance, particularly if the staggering tendency is persistently in one direction. As an example of the relative ease with which vertigo may be induced, it has been observed that a galvanic current passed through the head leads to a distinct tendency to fall toward the side of the positive pole, whereas a breaking of the current tends to induce falling toward the negative pole. That this phenomenon has direct relations with the semicircular canals is not to be questioned.

It is not possible to give a complete list of the causes and conditions which may induce vertigo. As a general statement, it may be said that anything which alters our normal spatial relations may be productive of vertigo. Some of the more important of these conditions are diplopia, rapid changes of position, especially in a rotary direction, looking from high places, and, more indirectly, through immediate or secondary involvement of the vestibular nerve, general increase of intracranial pressure, notably through tumors especially of the cerebellum, disturbances of circulation, particularly through arteriosclerosis, acute anemia, multiple sclerosis, migraine, epileptic attacks. Other causes may be attributed to disorders of the stomach, intestinal tract, nose, larynx, and to certain intoxications. Affections of the ear itself are naturally a prolific cause. Among the most commonplace and practically important of these is a collection of cerumen in the external auditory canal.

**Auditory Vertigo.**—In chronic middle-ear catarrh a type of disturbance may occur characterized at times by sufficient vertigo to cause a fall, accompanied with nausea, vomiting, and increase of tinnitus and deafness. Such attacks have been given the name Ménière's symptom-complex, to distinguish them from true Ménière's disease, which is presumably a labyrinthine affection, and which should in general be somewhat sharply distinguished from various other forms of vertigo which may under certain circumstances simulate it. So far as possible it is well to establish a pathological basis in the various forms of vertigo, and this seems possible in true Ménière's disease.

**Ménière's Disease.**—In 1861 a French physician drew attention to the group of symptoms which have since passed under his name. The terms auditory<sup>1</sup> and labyrinthine may be regarded as essentially synonymous with Ménière's disease. The words, at least, are used interchangeably. In limiting the application of this affection it is desirable to realize that it is through the immediate disturbance of the semicircular canals of the labyrinth that the symptom is produced. It should not, for example, be applied to vertigo manifestly due to distant conditions in the nervous system or organs which may, in a secondary way, affect the vestibular nerve. The onset of a typical attacks of auditory vertigo is, in general, as follows: It comes on suddenly, often sufficient to throw

<sup>1</sup> Dr. Clarence J. Blake has made a very useful distinction between "auditory vertigo," due to disease of the true auditory mechanism of the internal ear, and "aural vertigo," due to affections of other portions of the ear.



the patient to the ground. He is for a varying time stunned and possibly at times loses consciousness momentarily. The vertigo may be either objective or subjective in type. Vomiting and nausea quickly supervene, often persisting for hours, with headache. In these cases there is usually diminished hearing, with reduced bone conduction and almost always tinnitus. Occasionally nystagmus, on looking toward the affected side, diplopia, and facial palsy may supervene. The attacks vary widely in frequency, they may occur daily, weekly, monthly, or at times become practically constant (*Status Ménière*). In the earlier descriptions of the disease the onset was regarded as apoplectic in origin in a previously healthy ear. This view has been broadened to the extent of applying the term "aural vertigo" to cases with recognized ear disease.

*Pathological Anatomy.*—Considering the affection in its narrower aspect, the lesion may be regarded as a primary disturbance in the labyrinth, usually hemorrhage, occurring in the course of syphilis, leukemia, gout, and similar constitutional affections or from injury or from local inflammation. Disturbance in the function of the endolymph is causative of the ensuing symptoms. Deafness naturally results from a lesion of the cochlea, whereas the vertigo depends upon the invasion or disturbance of function of the semicircular canals. Parkes Weber, quoted by Osler, has made the following classification which is rather more general and inclusive than the limited conception of Ménière's disease as due only to disease of the labyrinth would justify. It is, however, of value as a comprehensive statement of the main causes of vertigo. "(1) The apoplectic form, due to hemorrhage into the labyrinth, as in leukemia, followed, as a rule, by complete deafness in one or both ears. (2) The cases associated with progressive inflammatory disease of the labyrinth. (3) Associated with organic changes in the auditory nerves, as in tumors, sometimes in tabes, and in cases of aural vertigo associated with facial paralysis on one side. (4) Cases in which a paroxysm of epilepsy is preceded by an auditory aura. (5) The moderate attacks which are associated with the various middle-ear affections, with wax in the meatus, with violent syringing of the ears, etc., all of which are probably due to increase in the intralabyrinthine pressure. Ménière's symptoms may occasionally be due to temporary excessive increase in the perilymph, possibly of angioneurotic character."

*Diagnosis.*—When occurring in typical form and when associated with manifest disturbance in the auditory function this is not difficult. In any condition of vertigo, the ear should be subjected to a careful and expert examination, to determine the range of hearing and the possible affection of the internal ear, even if the patient's attention has not been especially directed to it through tinnitus or other symptoms. The other conditions which may be indirectly the cause of vertiginous attacks should likewise be carefully investigated. It is not uncommon to attribute to a disordered stomach conditions which are due primarily to labyrinthine disturbance and of which the stomach disorder is an effect.

The greatest diagnostic difficulty lies in a sharp distinction between Ménière's disease and epileptic seizures. In the presence of marked labyrinthine disturbance manifested through deafness or tinnitus, the

differentiation should not be difficult. When these symptoms, however, are slight or possibly lacking, Ménière's disease may very closely simulate an epileptic attack. For example, aural vertigo may be and often is extremely sudden in onset, extremely short in duration, may unquestionably be accompanied by temporary loss of consciousness, may occur during sleep, and finally, which naturally renders its separation from epilepsy impossible, may be actually associated with that disease. All the foregoing conditions are met with in epileptic seizures in essentially the same form. Evidently, therefore, the points of distinction may be extremely vague, at least in exceptional cases. The violent sense of impulsion, as if being "hurled to the ground," is no doubt characteristic of aural vertigo, and does not occur in the same form in the epileptic seizure. Convulsive movements naturally do not occur in aural vertigo, nor is the onset so sudden that the patient is likely to injure himself. Gowers<sup>1</sup> has described in much detail the extraordinary difficulties which may sometimes arise in the differential diagnosis between these two affections. Stress should always be laid upon the existence of ear symptoms in a supposed Ménière complex. Such symptoms naturally may occur as a complication of a true epilepsy, but if epilepsy is excluded point strongly to the labyrinth as the primary source of the affection.

In certain debilitated states of the nervous system, symptoms closely resembling, if not identical with Ménière's disease, may occur. It has been suggested that under these conditions vasomotor disturbances in the labyrinth may actually lead to a true auditory vertigo.

*Prognosis.*—The outcome of Ménière's disease is always problematical. Experience has shown that with the increase of deafness the vertigo often decreases, and finally, when deafness is complete, may entirely disappear.

*Treatment.*—It is natural that in a condition of this sort a wide range of treatment has been suggested. From what has been said it is evident that attention should primarily be directed to the condition of the ear. It is found, however, that simpler methods of treatment, such as middle-ear inflation, often fail of definite result, making a resort to surgical intervention almost imperative. Eugene A. Crockett, of Boston, has drawn attention to the operation of removal of the stapes for the relief of vertigo. He regards this, however, as justifiable only in those cases in which ordinary treatment has proved futile. Less radical operations on the ossicles are suggested as preliminary measures of relief. The still more radical operation of removal of the semicircular canals necessitates the almost certain destruction of the cochlea as well, with resultant complete deafness. Even this operation, however, may in certain intractable cases be justified. The following general methods of treatment are at times serviceable: The bowel movements should be kept perfectly free, warm foot baths may be given, a residence at Karlsbad or Marienbad and similar watering places may be advised. Iodide of potash may be used to combat a possible syphilitic cause. Ocular and nasal defects should be corrected. Charcot advised quinine in spite of its known effect on the auditory nerve. The salicylates have also

<sup>1</sup> *The Borderland of Epilepsy*, 1907.

been prescribed and the bromides have undoubtedly been found useful at times. Nitroglycerin is indicated in high arterial tension associated with arteriosclerosis. Pilocarpine injections have been extensively used, 5 to 8 drops of a 2 per cent. solution every second day, often continued for weeks, or  $\frac{1}{8}$  grain subcutaneously or by the mouth. Lumbar puncture, advised by Babinski, in which from 3 to 20 cc. of fluid are withdrawn in one or several sittings, is a somewhat doubtful procedure.

**Paralyzing Vertigo (Gerlier's Disease).**—Although vertigo is but one of the symptoms of this extraordinary affection and its relation to the auditory nerve is not understood, nevertheless it may properly be discussed at this point. Gerlier,<sup>1</sup> a Swiss physician, first described the affection in 1887. In Europe the affection has been observed only in Collex, a canton of Geneva, and in certain surrounding hamlets. Cowherds and workers in the fields alone appear to be its victims, and they only during the summer months and under poor hygienic conditions. Vigorous young men are particularly likely to be attacked.

**Symptoms.**—Pain in the occipital region, muscular weakness, and disorders of the ocular nerves constitute the characteristic symptoms, combined in various ways and in varying degrees of intensity. Vertigo is a somewhat less conspicuous symptom. The pain in the neck is described as a sense of constriction often radiating into the back. The muscular weakness affects the voluntary muscles and particularly the extensors. The neck muscles may be unsymmetrically involved. An unequal ptosis is a common symptom not accompanied by paralysis of external or internal ocular muscles; the visual fields are at times diminished, but the fundus shows nothing characteristic. At the onset of an attack and preceding the ptosis a certain clouding of vision with vertigo often occurs. A typical attack may be described as follows: "The patient, previously well, is suddenly attacked by pain in the neck and back; his sight is clouded to the point of temporary blindness; ptosis develops rapidly along with general weakness of neck and body muscles; standing erect becomes difficult; he reels and has all the appearance of drunkenness. After a period not exceeding ten minutes complete recovery takes place. During an attack the tendon reflexes are preserved, at times increased; the skin sensibility is intact, false trismus occurs, swallowing is often impossible, and general motor weakness dominates the clinical picture. The head often falls forward on the chest, due to weakness of the neck muscles, which, with the ptosis cutting off vision, renders the patient for the time wholly helpless. Nausea does not occur. Many attacks are slight and merely momentarily incapacitate the patient, but the foregoing symptoms invariably are developed in some degree."

**Etiology.**—The cause remains undetermined. Considering the conditions under which the affection is most likely to occur, Gerlier presumed that it was due to an infection derived from stable soil and active only in the warm months of summer. The malady does not occur in cold weather, and the attacks are usually in the afternoon, ceasing shortly

<sup>1</sup> *Rev. méd. de la Suisse romande*, 1887, vii, 1, 260; *ibid.*, 1888, viii, 22, 86; *ibid.*, 1891, xi, 201, 260.

after sunset. Other explanations which have been offered, including hysteria, are even less satisfactory than Gerlier's suggestion. Although this type of vertigo, so far as known, occurs nowhere else on the Western Continent, a similar affection was described by Miura in the northern portion of Japan, to which was given the name "Kubisagari."

*Treatment.*—Improved hygiene appears to be the most rational form of treatment. The affection is never fatal, and occurs only in summer.

### GLOSSOPHARYNGEAL (NINTH) NERVE.

**Symptoms.**—Isolated disease of this nerve is not definitely known, or at least the cases are too few upon which to base conclusions of value. It follows, therefore, that exact knowledge of its function is vaguer than that of any of the other cranial nerves. Clinical investigation and experiment have established beyond much doubt that the dorsal portion of the tongue and neighboring parts of the pharynx are supplied with taste fibres through it. The question has, however, been raised that possibly even these fibres finally reach the brain through the fifth nerve. Others are of the opinion that the chorda tympani fibres finally enter the central glossopharyngeal. This and similar disputed points are difficult of determination on account of the rarity of affection of this nerve without coincident involvement of its neighbors. Cassirer, working with Oppenheim, has reported a case which, in their opinion, demonstrates the fact that in certain instances all the taste fibres may run in the glossopharyngeal. In addition to the innervation through the glossopharyngeal, parts of the posterior portion of the tongue and neighboring parts are also innervated by the fifth nerve, which adds still further to the difficulty of exact determination of the boundaries of the two sensory supplies. It may, however, be accepted that the chief innervation of the pharynx is from the glossopharyngeal. So far as this nerve is concerned with the act of swallowing and with the motor innervation of the œsophagus in general, its relations to the vagus presumably must be held responsible. Its direct relation to salivary secretion is probable. The nerve is very frequently involved in disease of the bulb, but rarely alone, as would naturally be inferred from its extremely close central relations, particularly with the vagus and accessory nerves. Disease external to the bulb in the posterior portion of the skull, aneurism, tumors, thrombosis of the jugular vein, may implicate the nerve, but here again rarely alone, nor are injuries any more likely to affect it and not others. Oppenheim describes an interesting case following protargol insufflation, in which the symptoms were slight paralysis of the palate and throat muscles, with loss of taste in the parts supplied by the nerve. This condition he thought due to chemical action on the nerve endings. Degeneration of the nerve may occur in tabes. Disturbances of taste in affections of the middle ear due to involvement of the tympanic plexus and increase of salivary secretion have been observed.

**PNEUMOGASTRIC (VAGUS, TENTH) NERVE.**

**Etiology.**—Although the vagus nerve is seldom affected by a primary isolated neuritis, it is easily involved through other causes because of its extremely long course and wide distribution. There is evidence to show that the nerve may take part in a general neuritis or analogous conditions. For example, the nerve may be affected in a general alcoholic neuritis, and its involvement in diphtheria, as described by J. J. Thomas<sup>1</sup> and others, is recognized as possibly one of the causes of sudden death in that disease. The nerve may also be affected in other infectious diseases and also in intoxications through lead and arsenic. Tumor, aneurism of the vertebral artery, meningitis, and similar disturbances external to the oblongata may injure the nerve, but in this case, as with the glossopharyngeal, neighboring nerves are usually affected at the same time, among which the ninth, eleventh, and twelfth are the most frequent. Sclerosis of the vessels of the posterior fossa may lead to disturbance in the function of the nerve, and it is not infrequently injured in operations for tumors of the neck and in tying the carotid artery. Conditions affecting the bulb itself, as, for example, softening, hemorrhage, bulbar paralysis in its various forms, tabes, and multiple sclerosis, are responsible for lesions of central origin. A noteworthy lesion is the involvement of the nerve in tabes, which presumably leads to the crises characteristic of that disease. In many of the hysterical conditions on the part of the gastro-intestinal, respiratory, and circulatory systems the nerve plays an important part.

**Symptoms.**—The symptoms produced by affections of the vagus nerve are naturally varied because of the possibility of wide location of the lesion producing them. A total paralysis of the nerve is rare, but may occur through extensive destructive processes in the posterior fossa. In this case, however, lesions of other nerves are a usual accompaniment as already suggested. In one-sided paralysis the symptoms of unilateral weakness of the palate and larynx are conspicuous. The soft palate is flaccid and hangs away from the affected side, not moving perfectly with phonation. Speech is nasal and there is more or less difficulty in swallowing. The vocal cord on the affected side does not take part in phonation or respiration. There may also be partial anesthesia of the pharynx and larynx. In unilateral lesions of the nerve the condition of the heart is not constant. At times there is slowing of the beat, but more often an increase. Respiratory disturbance occurs usually only with a double lesion. Other general symptoms due to irritation or paralysis of the vagus are vomiting, bulimia, loss of appetite and thirst, pain in the epigastrium, and, conceivably, production of a diabetes.

**Pharyngeal Branches.**—The innervation of the pharynx is through the vagus and pharyngeal nerves by way of the pharyngeal plexus. It is difficult to determine the exact part played by each nerve in the paralyses. The symptoms of disturbance in this field of innervation are difficulty in swallowing, inasmuch as the food is not properly passed on or enters

<sup>1</sup> *Med. and Surg. Reports*, Boston City Hospital, Ninth Series, 1898, p. 52.

the larynx, or, if the soft palate be involved, passes into the posterior nares. The so-called nasal voice is a common accompaniment. This group of symptoms is characteristic of lesions of the nuclei of origin, as seen in bulbar paralysis, in which death commonly results from actual starvation. The involvement of the pharynx with good prognosis is also common in postdiphtheritic paralysis. The difficulty in swallowing is very slight or negligible in one-sided paralysis. Spasmodic affections of the pharynx often occur without known structural basis, and are at times important as part of a more general neurosis, as, for example, hysteria. A pharyngeal spasm may at times be induced by exaggerated self-consciousness. It occurs also as one of the symptoms of hydrophobia.

**Laryngeal Branches.**—The importance of a laryngeal examination in all cases where there is a suspicion of the involvement of this region cannot be exaggerated. A palsy of the larynx may exist entirely without objective symptoms. For example, in common abductor paralysis there is not necessarily any disturbance in vocalization, but the quality of the voice is usually slightly changed.

The innervation of the larynx is through the superior and inferior or recurrent laryngeal nerves. The superior laryngeal nerve supplies sensation to the larynx and motor fibres to one pair of muscles, the cricothyroids, which act as tensors by separating the points of attachment of the vocal cords on the thyroid cartilage from those on the arytenoids. Paralysis of this nerve, therefore, leads to relaxation of the cord or cords, which, therefore, become slack on phonation. The voice is not lost, but is weak and easily tired. The paralysis is not well marked. Paralysis of the superior laryngeal nerve is rare because of its short course, but may be produced by wounds, tumors, or enlarged glands. It is also often obscured by the much commoner paralysis of the muscles of the cords.

The other muscles of the pharynx proper are wholly supplied by the recurrent laryngeal nerves, which also give sensory fibres to the mucous membrane below the cords. The left recurrent nerve curves around the arch of the aorta and the right passes under the subclavian artery. It is natural, therefore, that these nerves are peculiarly liable to pressure from aneurism and also on account of their long course through involvement by tumors, glands, and other processes in the upper thoracic region. Varied palsies of the laryngeal muscles may result, giving rise to a wide diversity of symptoms and to a varied laryngeal picture.

In conditions of health the edges of the vocal cords are straight on phonation, parallel and nearly in contact. Abduction takes place on deep inspiration and there is slight movement in quiet breathing. Resting, the glottis is wider than when in the cadaveric position or in complete paralysis, due to the slight tonic action of the abductors. In gradual degeneration of the recurrent laryngeal nerves the paralysis of the muscles follows a definite order (Semon's law). Abduction is first affected through disturbed action of the posterior crico-arytenoids. The thyro-arytenoids are next involved, and then the lateral crico-arytenoids. The arytenoid, unpaired, has a bilateral nerve supply, and is, therefore, not affected in unilateral cases. The analogy in this law to paralysis of skeletal muscles of the arm and leg is interesting.

**Abductor Paralysis.**—In unilateral abductor paralysis the affected cord lies in the middle line. On phonation the sound cord abducts and the larynx looks normal. The palsy becomes evident on inspiration. The voice may not be affected. Owing to the reduction of the aperture between the cords, dyspnoea occurs on exertion, but in adults ordinary quiet breathing is not affected. The symptoms of dyspnoea are more marked in children, owing to the relatively small size of the glottis. In bilateral abductor paralysis both cords lie near the middle line. On inspiration, owing to the shape of the cords, they act as a valve and are drawn closer together, causing marked inspiratory dyspnoea with stridor. In this form of paralysis sudden death is always possible from a cutting off of the air current. On failure of the thyro-arytenoids, the cords are no longer tense and hoarseness ensues.

**Total Unilateral Recurrent Paralysis.**—In this type of paralysis the cord lies in a position between abduction and adduction, the so-called cadaveric position. On inspiration the sound arytenoid is abducted and lies behind the other. On phonation the sound cord moves across the middle line, and lies at a slightly higher level than the other. The voice is not lost but is hoarse and easily tired.

**Bilateral Recurrent Paralysis.**—The severe dyspnoea which occurs in bilateral abductor paralysis does not occur when the paralysis is more complete, but the voice is completely lost. Both the cords lie in the cadaveric position, but slight inward movement from the partially active arytenoid may occur, presumably through its possible supply from the superior laryngeal nerve.

**Diagnosis.**—The diagnosis of these various forms of paralysis is made chiefly from the laryngoscopic appearances and from conditions of phonation and respiration.

**Functions of the Larynx.**—There are two main functions of the larynx—(1) phonation, and (2) respiration. The adductors are essential to the former and the abductors to the proper performance of the latter. *Phonation*, being a later acquired function, is under direct cerebral control, the centres controlling it presumably lying in the lower part of the precentral area, with further connections with the areas presiding over the general function of speech. Stimulation of the cortical centre for phonation in either hemisphere leads to adduction of both cords, demonstrating, as is usually the case in muscles which act under normal circumstances together, that the innervation is not entirely from one hemisphere. *Respiration*, which is the more fundamental function, and which in a certain sense is antagonistic to phonation, persists after the destruction of both cerebral hemispheres, as experimentally demonstrated. These facts are of interest in connection with the observation that so-called functional disorders lead to paralysis of the adductors, whereas abductor paralysis is due to structural changes. In lesions lying above the bulbar nuclei, if unilateral, paralysis of the cords does not ensue, as, for example, in hemiplegia, even though it be associated with aphasia. A bilateral lesion of the central tract, on the other hand, causes an adductor paralysis. Direct paralysis of the recurrent laryngeal nerve may result from lesions in the oblongata, at the base of the brain, in the vagus nerve, or in the

recurrent laryngeal itself. In nuclear disease affection of this nerve is usually combined with that of others, often associated with a persistent rapidity of the pulse. Syphilis, meningitis, or other disturbances in the posterior fossa may lead to laryngeal paralysis. A primary neuritis analogous to that of the seventh nerve is unusual, but has been described; paralysis from toxic agents is far less unusual. The vagus nerve in the neck may be variously injured. The frequency of laryngeal difficulty in the course of tabes, although not definitely understood, is of interest particularly in relation to the laryngeal crises. Anesthesia and various paresthesias of the larynx are not infrequent in this affection.

The condition of the larynx in tabes has been carefully studied by Dorendorf, of Berlin, in 1903, and by D. Crosby Greene,<sup>1</sup> of Boston, who found in 60 cases of tabes that 15 per cent. showed laryngeal complications, 10 per cent. paralysis of one or both vocal cords, and 12 per cent. were affected with laryngeal crises, figures which differ rather widely from those secured by Dorendorf on a basis of 245 cases. The type of paralysis in Greene's cases was always abductor, and this much more often unilateral than bilateral. The reason given for this fact is that the lesion is presumably a progressive degeneration of the fibres of the recurrent laryngeal nerve, and inasmuch as the abductors are the weaker group, they are the first to succumb. Greene further found that laryngeal crises are among the earliest laryngeal manifestations of the disease, from which the general practical conclusion is drawn that tabes as an etiological factor of certain laryngeal difficulties should be carefully considered. In certain cases, undoubtedly, the diagnosis of the more generalized disease may be made from the laryngeal signs.

Perhaps the most usual causes of the involvement of the nerve are the trauma of surgical operation, the presence of aneurism and enlarged glands, cancer, goitre, effusions in the pericardium, and mediastinal disease. The involvement of the recurrent nerve is unilateral or bilateral, depending upon the extent of the causative process. Aneurism is, no doubt, the most important single cause, but tuberculosis is also often operative. Involvement of the right cord should, in fact, suggest the possibility of tuberculosis of the lung, because of the fact that the nerve in its course downward lies close to the pleura and may easily be involved.

**Treatment.**—This comes almost wholly within the province of the laryngologist. The practitioner, however, is often called upon to treat the general condition, of which a laryngeal affection is one of the symptoms. Constitutional disease, for example, such as syphilis and tuberculosis or a general neuritis, must be treated by appropriate means irrespective of the laryngeal condition. The application of faradism to the affected laryngeal muscle under guidance from the laryngoscope is, at best, a doubtful procedure, and would certainly not be undertaken by any but a specialist. Surgical intervention in the various conditions which may lead to compression of the nerve in the neck and mediastinum falls within the province of the surgeon.

<sup>1</sup> *Boston Med. and Surg. Jour.*, 1906, clii, 97.



**Adductor Paralysis.**—As has before been said, adductor paralysis leading to difficulty or abolition of phonation forms a distinct class of functional paralysis. The great part which *hysteria* plays in these paralyzes has been a subject of much discussion. It is probable, however, as pointed out by Cartaz, Janet, and others, that in hysterical conditions there is not much disturbance in the vocal cords, and that, as a matter of fact, the paralysis is apparent rather than real. In this, as in other hysterical paralytic conditions, the source of the paralysis is unquestionably central, and lies rather in a central incapacity for proper innervation than in any disorder of the cords themselves. In other words, in hysterical mutism, if the patient were able to utter the sound which would lead to the proper action of the cords, the cords would no doubt functionate properly. The movement of the cords, however, cannot be made simply because the words or proper sounds cannot be framed, and this naturally is purely a mental process. In general, it is probable that the so-called hysterical origin of adductor paralyzes has been exaggerated, inasmuch as such disturbances are very frequently due to local disease immediately affecting the proper use of the laryngeal muscles. It may be said that adductor paralysis is never due to organic disease in the passage of the nerve from the bulbar nuclei to the larynx, but is either a pure neurosis or due to local disease. These myopathic paralyzes in some degree are frequent in various forms of laryngitis. Although aphonia may be and often is a manifestation of hysteria, most of the cases are not purely hysterical in the proper sense of that term. Any state, for example, which requires greater effort than normal for proper phonation, as, for example, debilitated conditions in general or laryngeal catarrh, naturally predisposes thereto. Difficulty in phonation is especially common in tuberculosis on account of the frequent combination of extreme debility and laryngitis apart from immediate involvement of the nerve. In hysterical conditions whispering is usually possible, but even this may be at times lost, leading to the condition of so-called hysterical mutism.

Adductor paralysis is almost always bilateral and seldom complete. It usually consists merely in an imperfect apposition of the cords. There are three sets of adductor muscles—the lateral crico-arytenoids, the thyro-arytenoids, and the unpaired arytenoid. These muscles may be affected together or irregularly.

**Diagnosis.**—Adductor paralysis may be mistaken for total recurrent paralysis. In neither do the cords come together on phonation. In incomplete adductor palsy the cords separate widely on deep inspiration, whereas in complete paralysis of the adductors the cords lie in a position of extreme abduction, which should prevent confusion of interpretation.

**Treatment.**—This falls within the special province of the laryngologist except when the affection is due to a general neurosis. The neurologist, however, is often called upon to treat the aphonias and mutisms which depend wholly or in part upon mental, hysterical causes. Suggestion variously applied, particularly by means of a strong faradic current applied over the larynx, will frequently restore speech. Such superficial methods of treatment, however, are usually temporary in effect and

relapses are the rule. A much more rational treatment is a painstaking investigation of the past life of the patient, with particular reference to the cause or causes which may have induced the speech defect.

**Spasm of the Laryngeal Muscles.**—This leads to adduction of the cords, as, for example, artificially induced by stimulation of the cortex. The abductors, if affected, are overpowered by the stronger adductors. Such spasmodic affections are usually bilateral, even if due to irritation of the vagus fibres on one side. The causes are irritations of the nerve in its central or peripheral course, and naturally precede the paralytic conditions due to a destruction of the nerve. Therefore, any pressure of mild degree on the nerve in its peripheral course, tumors in early stages, slowly growing aneurism, and local disease of similar sort may lead to the condition of spasm. A structural central lesion may likewise be a cause, and the condition has been observed in tabes, general paralysis and hydrophobia. In hysterical conditions, instead of a loss of function of the laryngeal muscle, there may likewise be an overaction leading to spasm. In children the condition known as "*laryngismus stridulus*" is of practical importance.

**Phonic Spasm (Spastic Dysphonia).**—This may be classified as a rare occupation neurosis allied to writer's cramp and similar affections. It occurs particularly in voice users, and gives rise to a variety of spasmodic affections similar to those just described.

**Sensory Neuroses.**—Anesthesia of the larynx, complete or partial, may result from injury of the nerve through disease, notably diphtheria, or from bulbar affections of varied character. Except in purely functional conditions, the motor portion of the nerve is ordinarily involved as well. The result is that food easily enters the larynx, often resulting in a fatal deglutition pneumonia. The *prognosis* is unfavorable in cases of total bilateral anesthesia. The *treatment* must be of the underlying condition so far as is possible. Special care must be taken in feeding the patient to prevent the passage of food into the larynx. In unilateral cases the patient should be directed to swallow very slowly, lying on the sound side during the process. In complete sensory paralysis the œsophageal tube is necessary, the strictest care being taken not to pass it into the insensitive larynx. Hyperesthesia and paresthesia of the larynx are not uncommon in certain neuroses and in tabes.

**Cardiac Branches.**—The cardiac plexus of nerves is formed from branches of the inferior laryngeal nerve from the main trunk of the vagus and from the sympathetic system. By these means the motor, sensory, and trophic functions of the heart are subserved. In one-sided lesions of the cardiac branches the symptoms on the part of the heart are not constant. At times there is slowing of the beat, but more often an increase in its rapidity. Such results have been observed both in experimental compression and in section of the nerve. In unilateral section of the vagus at times no symptoms whatever are produced, the laryngeal muscles even not showing the effect of the lesion. This no doubt is explained by the fact that muscles which ordinarily act bilaterally are innervated from both sides of the brain. The phenomenon at times observed of voluntary control of the heart is of interest in this connection. It

occasionally happens through some peculiar aptitude that a person is able, through an act of the will, to regulate within certain limits the rapidity of action of his heart.

**Sensory Symptoms.**—A consideration of the various cardiac neuroses dependent upon the disordered function of the vagus nerve would lead beyond the limits allotted to this section. It is one of the best recognized facts in medicine that the heart is peculiarly subject to functional disorders. Some of the conditions which may be attributed to a disorder of the sensory mechanism of the heart are palpitation, arrhythmia, tachycardia, bradycardia, angina pectoris and so-called pseudo-angina.

**Pulmonary Branches.**—Unless due to actual disease of the larynx or to organic disease of the lungs themselves, respiratory disorders are usually attributable to central causes or to bilateral lesions of the nerve. The evidence, however, thus far secured regarding the exact part played by the lesions of the nerve in the respiratory act is somewhat conflicting. Slowing, rapidity, and irregularity of respiration have all been observed. Oppenheim speaks of a case in which slowing of the pulse was associated with increase of respiration, attributable to vagus' disease. In tabes, a slowing of the respiration to three or four in a minute has been observed and has been supposed to be due to a double vagus lesion. A permanent disturbance of respiration does not occur in unilateral vagus disease. It is possible that the motor fibres controlling the bronchial vessels are concerned in the production of asthmatic conditions, and possibly also are responsible for a form of emphysema.

**Œsophageal and Gastric Branches.**—Branches of the nerve to the œsophagus are rarely diseased alone. A not infrequent condition, which must be classed among the functional disorders, is spasm of the œsophagus unassociated with organic stricture. A case under observation for many years in an elderly man without organic disease has shown marked improvement through treatment by faradism. In this instance, any carelessness in swallowing or any lack of proper mastication is liable to excite a violent spasmodic contraction of the lower end of the œsophagus, leading often to regurgitation and expulsion of food. The vagus nerve must be regarded as the sensory nerve for the stomach, through the local irritation of which the sensation of hunger is presumably induced. In its motor function to the stomach, its section lessens contractions. Vomiting is to be regarded as a sign of undue irritation, occasioned either by irritation from the presence of a foreign body, from overmuch food, or indirectly from the effect on the nerve of cerebral pressure. The gastric crisis in tabes is a phenomenon entirely analogous to the laryngeal crisis.

**Vagal Attacks.**—Under the heading "Vagal and Vasovagal Attacks," Gowers described seizures of peculiar type which he regards as due to disturbances of some of the functions of the pneumogastric. The symptoms of these seizures are, for the most part, sensory, and include subjective, gastric, respiratory, and cardiac discomfort, at times accompanied by cardiac pain and a sense of impending death. In addition to these symptoms there is frequently a slight accompanying mental change with vasomotor constriction of peripheral vessels. Slight tetanoid spasm and some sensory impairment complete the general clinical picture of

a somewhat vague but no doubt important symptom-complex. Seizures having the foregoing symptoms in varying degrees have a duration of ten minutes to one-half hour or more, occurring at varying intervals often for months or years. Gowers calls attention to the fact that a condition somewhat similar to these seizures has been described by Nothnagel and others, who have, however, not regarded the group of symptoms as features of a definite attack. The effect of compression of the vagus nerve in the neck, as shown by Waller many years ago, is, first, difficulty in breathing, followed by labored inspiration, uneasiness over the precordial region, and gastric discomfort at times amounting to nausea. If the pressure is continued, syncope results. These symptoms were not considered as due to compression of the carotid. In Gowers' experience women suffer more frequently from these vagal attacks than men, but he repudiates the idea that the seizures are to be regarded as manifestations of hysteria.<sup>1</sup>

### SPINAL ACCESSORY (ELEVENTH) NERVE.

The eleventh or spinal accessory nerve is purely motor in function. The portion of the nerve accessory to the vagus has already been considered. The spinal portion may be considered independently.

**Etiology.**—There are many conditions leading to involvement of the spinal accessory nerve and consequent disturbance in the functions of the two large muscles which it in part supplies. Lesions of the cervical cord itself, with destruction of the cells of origin of the nerve, are not infrequent. The more important of these are cervical myelitis, progressive muscular atrophy of spinal origin, gliosis and syringomyelia. Disease of the bone, notably tuberculous processes in the upper cervical vertebræ, pachymeningitis of tuberculous or syphilitic character, are likely to involve this nerve. Injuries of the nerve in the neck, particularly in operations for tumors, and of late the voluntary injury of the nerve to provide an anastomosis with the facial in cases of intractable facial paralysis, naturally lead to its partial or complete paralysis. A primary neuritis has also been described. In most instances, especially when the lesion lies outside the cord, the affection of the nerve is one-sided.

**Symptoms.**—The symptoms of paralysis are purely motor and consist in paralysis of the sternocleidomastoid and partial paralysis usually of the trapezius. Under ordinary conditions of rest a paralysis of one sternomastoid muscle does not lead to noticeable deformity. The defect, however, is brought out when the head, better against resistance, is turned toward the side away from the affected muscle, with slight raising of the chin. In paralysis of the right sternomastoid such a movement toward the left will be incomplete and no contraction of the

<sup>1</sup> "Women suffer more frequently, but these attacks are also met in men. This and the fact that the pneumogastric and vasomotor systems are readily influenced by emotion have probably led to the frequent submergence of these attacks beneath the vague conception of hysteria, a conception which conceals whatever it covers. We must rescue from it whatever we wish to study. It can be put back again afterward if desired."—*Borderland of Epilepsy*, p. 19.

muscle will take place. If the sternomastoid is paralyzed on both sides, the head tends to fall backward, and in a horizontal position cannot easily be raised. The attachments of the trapezius are such that a certain distinction may be made between the paralysis of its three portions. Under normal conditions the muscle, through its attachments in the neck and throughout the thoracic region of the spine to the upper and outer portion of the scapula, tends to raise the shoulder and approach the scapula to the middle line. The upper portion of the muscle from the occiput to the outer third of the clavicle draws the head backward and turns it slightly toward the opposite side. The middle portion from the ligamentum nuchæ and the three upper thoracic vertebræ to the acromion and outer portion of the spine of the scapula elevates the scapula. The lower portion from the fourth and subsequent thoracic vertebræ to the inner half of the spine of the scapula draws the scapula toward the middle line. In paralysis of the clavicular portion of the muscle the shoulder does not move in respiration, but the failure of backward motion of the head is inconspicuous because of the continued action of deep neck muscles. This upper bundle of the trapezius is very apt to be spared in paralytic conditions. Paralysis of the middle portion leads to a sinking of the acromion through lack of fixation, which the trapezius ordinarily gives it, and through the continued action of the levator anguli scapulæ. The shoulder is therefore depressed forward and cannot be raised. The raising of the arm suffers somewhat in consequence. Paralysis of the lower portion leads to an increased distance of the inner border of the scapula from the middle line, with increased prominence of the scapula as a whole.

**Prognosis.**—The prognosis of lesions of the spinal accessory nerve from the causes given above naturally depends upon their individual curability. In syphilitic conditions and those resulting from transient inflammations a favorable outlook may be anticipated. In general, if the nerve be given opportunity, it will rapidly regenerate without special treatment. In hopeless paralysis much comfort and improved action of unaffected muscles may be obtained by the application of apparatus to elevate the shoulder.

**Spasmodic Affections of the Neck Muscles.**—Spasm of the neck muscles, including the innervation of the spinal accessory nerve, is perhaps the most distressing form of this type of disturbance. The variety of spasm which may develop in this general group of muscles is so great that merely an artificial classification may be made. Although the sternomastoid and trapezius frequently take part in these spasmodic affections, and often predominate in the clinical picture, we are usually not justified in regarding the spasm as wholly under the control of the spinal accessory nerve, inasmuch as other muscles supplied from other sources are almost invariably involved. Among these other muscles, the splenius, scaleni, and deep muscles of the neck, and at times the platysma and omohyoid, are of particular importance. The fact should be recognized that, although for purposes of clinical convenience we regard certain common spasmodic affections of the neck as due to disturbance in the spinal accessory innervation, the affection is usually much more

widespread and shows a distinct tendency to pass to other muscles. The importance of this in relation to treatment will receive attention later.

**Torticollis.**—Torticollis, or wryneck, occurs in various forms, some of which are quite independent of the nervous system. Of these the following may be mentioned: Rheumatic torticollis, the ordinary stiff neck, has no relation with a true spasm. A fixed torticollis due to congenital or other shortening of the muscles supporting the head is likewise not a form of spasm. A type of acquired torticollis, usually of slight degree, which may lead to confusion in diagnosis, is that associated with carious processes in the upper vertebræ in the absence of external deformity. An abnormal and rigid position of the head, espe-

FIG. 35



Torticollis. Showing spasm of the right sternocleidomastoid muscle, and characteristic position of the head at the height of the spasm. (Massachusetts General Hospital.)

cially when associated with an otherwise unexplained exaggeration of the deep reflexes, should lead to the suspicion of a tuberculous or other process in the upper cervical region. The rigidity of the neck often seen in meningitis and other conditions which lead to pressure on the outgoing nerves is usually easy of diagnosis.

**Spasmodic Torticollis.**—The most practically important variety of spasm of the neck muscles is the so-called spasmodic torticollis, or wryneck, in which the muscles innervated by the spinal accessory nerve play a predominant but by no means exclusive part.

**Etiology.**—This is vaguely understood, hence a great variety of causes has been given, few of which are explanatory of the fundamental under-

lying condition which actually induces the spasm. Oppenheim, among others, is very strong in the conviction that the affection is observed particularly among persons with hereditary unstable nervous systems, although direct inheritance is exceedingly rare. Frequent association of torticollis with other nerve disorders, either neuroses or psychoses, lends a certain weight to this view, as does also the fact that many of the cases yield most satisfactorily to treatment by psychotherapy. To make the statement, however, that the cases occur primarily in an otherwise unstable nervous system does little to explain the exact nature of the mental or physical process which makes possible this special

FIG. 36



Spasmodic torticollis. Head drawn backward and chin up, due to involvement of the right trapezius, probably with certain deep neck muscles, in addition to the manifest spasm of the sternomastoid. (Massachusetts General Hospital.)

spasmodic manifestation. In view of the fact that there is no known pathological anatomy, we are entirely justified in assuming that the cerebral cortex plays an important part in the production of the spasm, whether or not it be on a previous neuropathic basis. Various somewhat fanciful explanations of the affection have been submitted which cannot be regarded as of universal application. Irritations in the distribution of the trigeminal and occipital nerves, localized peripheral irritations as from too small a collar, disorders in the symmetrical innervation of the two sides of the body, eye-strain, organic disease of the brain, intoxications of various sorts, even astigmatism, are some of the causes which have been assigned. Perhaps in the majority of cases no adequate exciting

cause whatever is to be found, and recourse must be had to the explanation that mental perturbation, overaction of the muscles, and habit are responsible.

*Symptoms.*—The form of spasm varies in different cases from slight, hardly noticeable involuntary movements of the head to violent spasmodic contractions, painful to the patient and extremely conspicuous to the onlooker. Clonic spasms usually associated with tonic muscular contractions are the characteristic forms in which the affection manifests itself. In those cases in which the spasm is clonic in character over long periods of time, one is naturally not justified in speaking of the torticollis as of the form here under consideration. The exact type of spasm depends upon the degree of involvement of individual muscles and upon the number and position of the muscles affected. The picture, therefore, becomes a very complex one, and it is often a matter of extreme difficulty to determine the precise extent of the muscular involvement. The common form and that most closely related to the spinal accessory nerve consists of a clonic and tonic spasm of the sternomastoid of one side, often accompanied by a similar spasm, particularly of the upper portion of the trapezius. In this type the chin and face are moved away from the affected side and the head bent slightly backward, due to contraction of the sternomastoid. The coincident contraction of the upper part of the trapezius likewise leads to a turning and backward bending of the head away from the affected muscle, thereby increasing in a general way and supplementing the action of the sternomastoid. Other forms of spasm, as, for example, of the splenius, rhomboids, and deep muscles of the neck, do not now concern us, particularly since they bear no immediate relation to the spinal accessory nerve. It is, however, very important to bear in mind that although a torticollis may begin with the spinal accessory innervation, it is likely to spread to other muscles. As suggested above, the extent and intensity of the spasm vary widely in different cases, and are unquestionably affected for the worse by unfavorable emotional conditions. Associated psychical disturbances are of interest particularly in their etiological relationship, but it should be borne in mind that a coincidental disturbance of a mental sort is perhaps particularly likely to arise in this distressing condition.

*Pathology.*—The attempt to locate the lesion productive of torticollis of the spasmodic type has been wholly unsuccessful. To say that it is a condition of irritation in the nerve nuclei is of small help, since we are ignorant of what such irritations may consist. The most reasonable hypothesis is that the cerebral cortex, particularly in the area concerned in the movements of the neck, is primarily at fault.

*Prognosis.*—The outcome is always uncertain, but never entirely hopeless. The unexpected not infrequently happens. In one instance, in which the condition had become so grave that an operation had practically been decided upon, a preliminary sea voyage afforded entire relief. In general, however, the affection is stubborn, resistant of treatment, and shows a tendency to spread to other groups of muscles, oftentimes with a substitution of a practically tonic spasm for the alternating clonic spasms of the earlier stages.



*Treatment.*—As in many affections the exact cause of which is not understood, treatment for spasmodic torticollis has been and remains largely empirical. It is not necessary, therefore, to enter into detail regarding the great number of drugs which have been advocated. Experience has unquestionably shown that, apart from their influence upon the nervous system in general, there is no agent that exerts the slightest specific action upon the spasm. The bromides and opium may be given, and as general sedatives may have an effect, but few would venture the prescription of the opium preparations unless as a last resort and under most unusual conditions. Drugs unquestionably play a very small part in the rational treatment of this affection. Electricity has been used in certain cases with good results. It has been suggested that in the application of the electrical current it is desirable to stimulate the sound muscles rather than those affected with the spasm in order to overcome, so far as possible, the overaction of the latter. However theoretically justified such treatment may be, it has relatively little practical significance. Hydrotherapy is, no doubt, useful as a general tonic measure, but it has no further significance.

The use of mechanical appliances is of interest, particularly from the fact that their effect in different cases varies widely. An arrangement by means of which the head is held absolutely fixed is never to be advised. Such constriction is extremely apt to excite the muscular spasm in an even greater degree, leading to a distressing situation for the patient, even although the head is mechanically prevented from moving. On the other hand, it has frequently been shown that a supporting apparatus which is suggestive in its operation rather than providing an absolute fixity, is often of great benefit. The common experience that in the midst of the most violent spasm the head may be restored to its normal position by gentle pressure applied to the chin is suggestive. The hint which these facts gives regarding rational treatment is that the mental side of the affection must always be seriously taken into account. The spasm is, in fact, semi-involuntary, at least in its earlier course, and should be treated accordingly. Following out this idea, the most useful treatment is certainly that which combines mental and physical means, and this is best attained through coördinative gymnastic movements. Such movements have as their underlying principle the possibility of substituting in a voluntary muscle a voluntary for an involuntary movement. The aim, therefore, is to educate the patient to again perform voluntarily movements over which he has lost control, or, otherwise expressed, to break up a bad neuromuscular habit. Systematic movements of the head are therefore advisable, and best carried out under the instruction of a skilled teacher. A purely psychotherapeutic treatment in these cases is, on the whole, not efficacious, nor is it necessary. Hypnotism is undesirable, certainly as a general procedure.

A wide range of surgical operations has been suggested and surgery would appear to be at first sight clearly indicated. Results have shown that surgical intervention is rarely curative and nearly always discouraging in its results. The reason for this is two-fold: first, that a surgical operation does not relieve the cerebral condition which undoubtedly

lies at the basis of the spasm; and, in the second place, that more than one muscle is apt to be affected, with a strong tendency to spread to still other muscles or groups of muscles. Simple resection of the spinal accessory nerve is of small value, owing to its rapid regrowth. Resection with evulsion is a more rational procedure. The operative procedure may go farther and resect upper cervical nerves if the increasing spasm demands. The muscles themselves may also be cut, thereby diminishing the strength of their contraction, and even more radical operations on nerves and muscles of the various structures about the neck may in rare instances be advisable. The results of operations have been such, however, that the unpleasant suggestion arises that the section of one nerve with cessation of spasm of the muscle to which it goes rather increases the tendency to spread. If the affection is primarily psychogenic in origin it necessarily follows that operation in itself is unlikely to effect a complete cure.

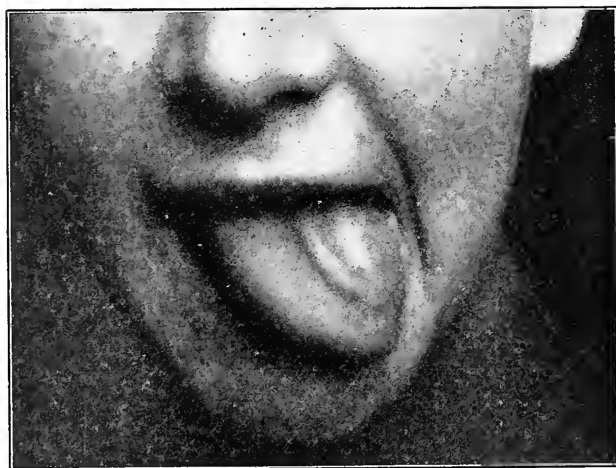
### **HYPOGLOSSAL (TWELFTH) NERVE.**

**Etiology.**—This nerve is very commonly affected in disease involving the bulb, notably in the various forms of bulbar paralysis, acute and chronic. In its central course from the lower portion of the cortical motor area to its secondary nucleus in the oblongata also it is not infrequently involved. Although not particularly noticeable on account of the action of the nerve of the opposite side, one hypoglossus is ordinarily affected in cerebral hemiplegia. This lesion, however, is less conspicuous, because unaccompanied with atrophic changes of the tongue. This fact, together with unchanged electrical reactions, is an important and simple means of differentiation between lesions of the central and peripheral neurone, being merely an example of a general principle. Lesions in the oblongata itself, however, naturally involve the cells of origin of the peripheral axones and lead to a characteristic atrophy of the tongue. Lesions external to the oblongata itself, as, for example, meningeal exudates, tumors, or hemorrhages at the base of the skull, may involve the nerve in its peripheral course, but usually in conjunction with others, and particularly the vagus. Aneurism of the vertebral artery and syphilitic processes may involve the nerve, but these are rare occurrences. As of other cranial nerves, an isolated neuritis is rare, although such a condition has been described by various writers. Congenital atrophies of the tongue, lingual hemiatrophy, and degenerations should also be mentioned in this connection.

**Symptoms.**—Disease of one or both hypoglossal nerves gives rise to signs not easily mistakable and of much diagnostic importance. These signs are perhaps particularly noticeable when the lesion involves but one nerve. As the tongue lies on the floor of the mouth there may be no deviation of its tip, but if moved, the tendency is for it to be projected toward the paralyzed side. The explanation of this constant phenomenon is not simple, owing to the complicated relations of the tongue muscles. The attempt to move the tongue within the mouth cavity is imperfect toward the diseased side, which naturally interferes somewhat with the

contact of the tongue on that side with various portions of the mouth cavity essential to proper enunciation and mastication. This defect, however, is so far compensated by the normal side that it leads to slight inconvenience. The defect becomes more apparent in the attempt to thrust out the tongue. This can be accomplished if but one side is paralyzed, but if the tongue is protruded the tip points strongly toward the affected side, its raphé describing a semicircle with its convexity toward the sound side; this deviation is due to the loss of contraction of the genioglossus. When the paralysis of both nerves is complete the tongue remains helpless and immovable in the mouth, and it becomes impossible for the patient to speak except in a most imperfect way. Mastication and deglutition are likewise interfered with. This condition is met with in the late stages of progressive bulbar paralysis. There is no sufficient evidence, as maintained by Gowers, that the muscles of the lips are innervated through the hypoglossal.

FIG. 37



Unilateral atrophy of the tongue. Protrusion toward the affected side, associated with movement of lower jaw toward the same side, owing to coincident involvement of the motor branch of the left fifth nerve. From a case of poliomyelitis.

*Atrophy* is the constant accompaniment of peripheral lesion of this as of other motor nerves. The tongue loses its normally smooth appearance, and is thrown into folds and ridges due to the wasting of the underlying muscle bundles. To the touch it is soft and non-resistant, shows alterations in electrical reaction, and is pervaded by a fibrillary tremor.

**Treatment.**—The treatment of paralysis of the tongue, either of the spastic or atrophic type, lies almost wholly in that of the underlying condition. Electricity applied to the atrophying tongue muscle is a practical impossibility. In many conditions of unilateral disturbance of the tongue the discomfort is not sufficient to call for treatment, even if it were possible, and in the sudden or gradual involvement of the nerve from bulbar or more peripherally lying disease the affection of the tongue

constitutes a relatively insignificant part of the clinical picture and demands no special treatment. The partial resection of the nerve which has lately been practised in faciohypoglossal anastomosis for facial paralysis leads naturally to immediate disturbance in its function characterized by a tendency to atrophy and slight speech defect, but this is quickly overcome, with complete restoration of function.

**Spasmodic Affections.**—The occurrence of spasm in the muscles supplied by the hypoglossal nerve is common in association with more general spasmodic affections, as, for example, epilepsy, hysteria, and chorea. The impossibility of controlling the tongue observed in general athetosis, often leading to a complete loss of power of articulate speech, is a somewhat analogous condition, but is not properly included under spasmodic affections. Much more rarely the tongue muscles are affected with spasm which either does not extend beyond them or only in the immediate neighborhood. Such spasms may be either tonic or clonic in character. If *tonic*, the tongue is apparently reduced in size and is pressed against the teeth or other parts of the mouth cavity. Under these conditions speech and swallowing are affected. Associated or independent of these tonic spasms a *clonic* condition of the tongue muscles may be developed. Under these conditions the tongue is rapidly thrust out or withdrawn, or makes various involuntary excursions within the mouth cavity. Such spasms may be either bilateral or unilateral in distribution, the latter being less frequent. The relation of these cramp forms to various speech defects known as stammering and stuttering is of interest. Under certain conditions the difficult speech is apparently induced by the incapacity of the person to move the tongue properly, with a result that the word or syllable is not enunciated, or in an explosive manner. However complicated the ultimate explanation of such forms of speech disturbance may be, the relation to spasmodic affections in the distribution of the hypoglossal nerve should always be borne in mind. The muscle spasm or tic is rarely constant, but occurs rather in the form of attacks, varying widely in frequency and not always ceasing during sleep. The causes are somewhat vague, but may in general be attributed to sources of irritation in the neighborhood, on the basis of a neuropathic tendency. The relation of the mental state to cramp attacks is undoubtedly important. They may, for example, be aroused by emotional disturbances of one or another sort. The *prognosis* is, on the whole, favorable, even though the tendency to spasm may have existed for years. The *treatment* consists essentially in the removal of any exciting cause and psychotherapeutic procedures, with such muscle training as is possible. Drugs and electricity play no essential part in the treatment.

## CHAPTER XIV.

### DISEASES OF THE PERIPHERAL NERVES.

By GORDON M. HOLMES, M.D., F.R.C.P.

THE symptoms of disease of the peripheral nerves may be classified as they result from disturbance of the functions of those fibres which convey impulses to the muscles, of those which carry the sensory impressions centralward, and finally, of the fibres which are more immediately concerned with the nutrition of the tissues, either directly or as part of the mechanism for the local regulation of the blood stream. There are, however, but few nerves in the body, excepting the cranial nerves, which are either entirely sensory or entirely motor. The consequence is that when any nerve is severely injured, both sensory and motor symptoms and probably, in addition, nutritive or trophic changes result. It is by the extent and distribution of these symptoms that the localization of the disease can be determined. If, however, the lesion or the disease of a mixed nerve is partial or incomplete, the functions of the different sets of fibres may suffer unequally; the general experience is that in incomplete lesions, as those due to compression, the sensory suffer much less than the motor fibres, or sensory symptoms may be absent, although the muscles supplied by the nerve are completely paralyzed. Lüderitz showed experimentally that the conductivity of the motor fibres is lost earlier than that of the sensory fibres when a mixed nerve is subjected to slowly increasing pressure. Evidence of trophic or vasomotor disturbance is, as a rule, little apparent in partial lesions.

**Motor Symptoms.**—The symptoms which immediately follow the complete interruption of the motor nerve fibres that supply a muscle are complete paralysis of both reflex and volitional contraction of that muscle, loss of its tone, and later, atrophy, changes in the character of its response to electrical stimulation, and finally, if recovery does not take place, contracture owing to secondary fibrosis. These features distinguish lower motor neurone or spino-muscular paralysis from paralysis due to disease in the upper motor or cerebro-spinal neurones. In the latter condition movements, not muscles as such, are paralyzed, and the distribution of the paralysis does not correspond to the distribution of one or more peripheral nerves; secondly, the tone of the paralyzed muscles is increased, not diminished or lost, that is, the paralysis is spastic and not flaccid, and the reflexes which are dependent on the tone of the muscle, as the knee-jerk, are more active than normal instead of being abolished; thirdly, the paralyzed muscles do not atrophy, or atrophy only to a much less degree; and finally, there is no change in the nature of their response to electrical stimulation.

The amount of *loss of power* in the muscles naturally depends on the degree of the injury of the nerve; when there is complete interruption of its structure or loss of its function the paralysis is necessarily complete. When the lesion of the nerve is incomplete another factor must be considered, namely, the rate of its evolution, as when the disease is of sudden or rapid onset the symptoms may be considerably greater, for a time at least, than those produced by a similar lesion which has developed slowly. A muscle that receives its motor fibres from more than one nerve is not completely paralyzed by even a complete lesion of one of them.

The *normal tone or tension of muscles* is dependent on the integrity of the peripheral reflex arc, which consists of the afferent fibres from the muscle that enter the cord by the dorsal spinal roots and terminate by synapsis around the cells of the corresponding motor neurones in the ventral horn, and of the peripheral motor neurones. When this arc is broken in any place the muscles immediately lose their tone. The atonia may be recognized by loss of the normal contour of the muscles, if this is easily visible; by their softness and flabbiness to touch, and by the lack or diminution of the resistance which they normally offer to stretching, and consequently the excess of mobility to passive movement of the joint at which they act.

The *deep reflexes or tendon-jerks* are dependent on the maintenance of the muscle tone, and it is by these so-called reflexes that this variety of tone in the muscles is most easily measured. Their disappearance or abolition is consequently an indication of the diminution or disappearance of the tone of the muscles concerned and not of their paralysis alone. The knee-jerks may disappear early in the toxic affections of the peripheral nerves, before there is any demonstrable evidence of either motor or sensory paralysis; and during recovery from such conditions, or after regeneration of degenerated nerves, they may be absent when the recovery of power is apparently complete. The diminution or loss of tone as measured by the deep reflexes is the most sensitive guide to any interference with the normal function of the peripheral nervous system.

*Atrophy of the muscles* supplied by the affected motor fibres is one of the most prominent symptoms; this is due to degeneration or regression of the muscle fibres owing to loss of the trophic influence which the motor nerves normally exert on them. It is recognizable by a diminution in the size of the muscle, and by its soft and structureless consistence. The latter symptom is frequently neglected, but it may prove valuable, especially in children, in whom it is not easy to determine the distribution of an atrophic paralysis by merely noting the movements which cannot be performed; here by touch alone we may pick out the affected muscles. Muscular atrophy is generally recognizable within two or three weeks after a complete interruption of a motor nerve, and increases rapidly from this time until few if any fibres remain. When the nerve lesion is incomplete the atrophy corresponds closely with the degree of paralysis; in other words, the conducting power and trophic functions of the peripheral motor nerves are lost together. When the lesion of the nerve is very slowly progressive the muscular atrophy is generally less prominent than the paralysis.

Coincident with the atrophy of the fibres of the wasting muscle an increase and proliferation of its connective tissue occur. If the muscular atrophy attains severe degrees, and if regeneration does not soon set in, this new connective tissue slowly undergoes fibrosis and the muscle is converted into a firm, inelastic band. The contracting fibrous tissue may produce contractures and deformities. Further, for the recovery of function it is necessary that the muscle fibres should regenerate, and regeneration must be evidently seriously interfered with by the presence of dense and contracted fibrotic tissue. The condition of the muscles and the changes they undergo are therefore as important as the changes in the affected nerves.

One of the most valuable signs in disease of a peripheral motor nerve is *change in the electrical excitability of the nerve and of the muscles it supplies*. Muscular contraction can be normally obtained by either faradic or galvanic stimulation of the motor nerve fibres which supply it. Two or three days after section of a nerve the excitability of its peripheral portion is diminished, and after a period of six to eight days the excitability is lost with the secondary degeneration of the nerve. The most characteristic changes are observed on direct stimulation of the muscle; they compose what is known as the *reaction of degeneration* (R. D.) and usually appear from the eighth or tenth day after section of the nerve. It must be mentioned that according to Sherren<sup>1</sup> increased excitability to the galvanic current is found only when the lesion of the nerve is incomplete. After some months, if recovery has not set in, the reaction of the muscles to the galvanic current diminishes slowly and finally disappears.

In every case of peripheral paralysis it is important to test the electrical excitability of every portion of the nerve and muscle that can be reached. It occasionally happens when there is a local lesion in the nerve that muscular contractions can be obtained by stimulation below the lesion, but not from above it; the lesion may block the conduction of impulses without leading to secondary degeneration of the peripheral portion of the fibre. On the other hand, regenerating nerves may be excitable only from above the lesion; the regenerated portion is for a time inexcitable to stimuli which it can conduct. This may depend on the absence of a myelin sheath (Erb). When the lesion is unilateral the excitability of both nerve and muscle may be compared with those of the normal side; when both sides are paralyzed the irritability should be compared with those of a normal person, or the tables prepared by Stintzing to show the normal limits of excitability may be used.

Ghilarducci's *distal reaction* is also a valuable test; the indifferent electrode is placed on the neck or back, the other distal to the insertion of the degenerated muscle; then on closure of the current a contraction of this is obtained by a weaker current than suffices for the unaffected muscles.

*Symptoms of irritation of motor fibres* are much less frequently met with in diseases of the peripheral than of the central nervous system. Spasm, either tonic or clonic, and cramps, when due to irritation of the peripheral

<sup>1</sup> Injuries of Nerves, London, 1908.

neurones, are generally reflex in origin and arise from excitation of the sensory fibres. Occasionally, however, intermittent or tonic spasm may result from irritation of a motor nerve by a neighboring focus of inflammation or by a foreign body, but even here it is difficult to exclude its reflex origin. True reflex spasms are much more frequent; the involuntary facial movements which often accompany trigeminal neuralgia, and even irritation of the cornea, are of this nature. The fixation of painful joints by the tonic contraction of the surrounding muscles is also due to irritation of the sensory fibres which reflexly excite the corresponding motor neurones to excessive tonic activity; the sensory impulses may, if sufficiently intense, spread to wider reflex centres in the cord. The potency of sensory impulses from the periphery in the causation of spasm is illustrated by the severe spasms met with in strychnine poisoning and in tetanus; both these poisons only transmute an inhibitory effect into an excitation effect in the spinal reflex centres (Sherrington), and the spasms or convulsions are always directly due to a stimulus from the periphery. Cramp of a muscle may be occasionally due to influences that affect its fibres directly, as poisons like veratrine; venosity of the blood may also predispose to or cause spasms.

Some forms of muscular atrophy are characterized by the occurrence of fibrillation or intermittent, more or less rhythmical wave-like contractions of some of the fibres of a muscle; this is more probably due to direct stimulation of the fibres than to excitation of the motor nerves. Myokymia, a condition characterized by constant undulating or wave-like contractions of the muscle fibres which changes from place to place, has been regarded as a symptom of abortive neuritis, but is rarely seen.

After complete division of a nerve and primary suture the time necessary for the return of power depends on the distance of the lesion from the periphery; Sherren lays down the rule for the upper extremities, that when a nerve is divided at the wrist perfect power may be regained within a year; but if at the elbow or in the plexus, not for two years. The muscles nearer the lesion regain their functions earlier than those distant from it. Recovery is slower after secondary than after primary suture. The muscular functions return much more rapidly after incomplete nerve lesions.

**Sensory Symptoms.**—When a sensory nerve or the sensory fibres of a mixed nerve are cut across we might expect to find absence of all sensation in the cutaneous and deep structures which are supplied by that nerve; but by the ordinary methods of testing this is rarely found, as the sensory loss which is revealed is generally much less extensive than the anatomical distribution of the nerve. This has been generally explained by assuming that the nerves overlap or anastomose, or that the neighboring end organs take up sensibility from the anesthetic region. But the explanation has been afforded by the brilliant work of Head and his colleagues.<sup>1</sup> These have shown that the afferent fibres may be divided into three systems. The first system subserves *deep sensibility*, which is conveyed, as Sherrington<sup>2</sup> has shown, by the afferent fibres that run from the muscles,

<sup>1</sup> *Brain*, 1905, xxviii, 99.

<sup>2</sup> *Phil. Trans. Royal Society*, 1896.



tendons, and joints, and which escape when only cutaneous sensory nerves are injured. Even when the muscular branches are involved this form of sensibility may not be quite abolished, as its fibres have wide anastomoses and often join the tendons and muscles high up in the limbs. Its function is the appreciation of pressure, of stimuli that produce deformation of structure, and of any change in the position and condition of the joints and muscles. It is the system which subserves the sense of position. It is owing to the persistence of deep sensibility that the statement is often made that no sensory loss ensues on the section of a cutaneous nerve, as the area of its distribution remains sensitive to even light pressure by a finger or pencil point.

The second system, to which the name *protopathic* is given, conveys painful cutaneous stimuli and the appreciation of the major degrees of temperature. But in the absence of the third system, the pain produced by a pin-prick or other means is not localized, but radiates widely over the affected area and causes an unnatural amount of discomfort and an almost uncontrollable desire to withdraw the part from the stimulus. Although it is through this system that major degrees of heat and cold are appreciated, minor degrees of temperature cannot be recognized when it is alone present, and the appreciation of degrees of heat and cold is lost. The third system, which has been called *epicritic*, responds to light touches and to the minor degrees of temperature, which produce the sensations called "warm" and "cool." It is only when epicritic sensibility is present that the point of skin touched can be accurately localized, and that two points at a normal distance apart can be discriminated when applied simultaneously.

In the sensory disturbances that result from section of a peripheral nerve the different varieties of sensibility are lost according to their arrangement in these three systems, that is, sensibility to light touch and the minor degrees of temperature disappear together, and the appreciation of the major degrees of temperature with insensibility to painful cutaneous stimuli.

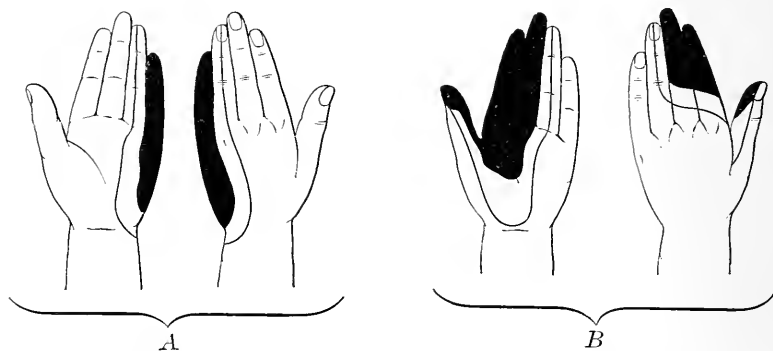
When the condition of sensation on the hand is examined after section of the ulnar nerve it will be seen that there is complete loss of all forms of sensation only on the little finger and on a variable, but small, area on the ulnar border of the hand; that is, it is only here that both the protopathic and epicritic sensibilities are absent (Fig. 38), but on the rest of the cutaneous distribution of the ulnar nerve light touch and the intermediate degrees of temperature, the epicritic sensibilities, cannot be appreciated, and although painful stimuli may be recognized they cannot be accurately localized, but radiate widely and give rise to unnatural discomfort. It is evident that though there may be a considerable overlap between the fibres of the ulnar and median nerves which conduct protopathic sensibility, the loss of epicritic is practically limited by the anatomical boundary of the nerve distribution. With other nerves there may be an overlapping of epicritic sensation too. When the fibres are injured in a plexus the area of loss of protopathic sensibility may almost equal in extent that of the epicritic, while if due to lesion of the dorsal spinal roots, the area insensitive to a pin-prick may actually

exceed that insensitive to light touch. The more closely a peripheral nerve represents the supply of one or more posterior roots the more nearly will the loss of protopathic coincide in distribution with the loss of epicritic sensibility. Head's conclusions have been contested by Trotter and Davies<sup>1</sup> and others.

When a nerve is not completely divided, or if only its functional continuity is affected, as by bruising or compression, the condition of sensation is very variable. Sometimes every form of sensibility is lost for a time, but often pain perception remains intact and loss of epicritic sensibility may be the only sign of the injury; or after slight injuries there may be no absolute loss of sensation, though the patient may be conscious of an altered sensibility in the area of the nerve.

The *recovery of sensibility* in the area of a divided nerve which has been sutured and is regenerating takes place in a definite and constant manner. The first change, generally observed after about two to three

FIG. 38



A, loss of sensation produced by division of the ulnar nerve; B, that due to division of the median nerve. The areas of complete cutaneous insensibility are marked black; the skin insensitive to light touch and the intermediate degrees of temperature are enclosed by the line. (After Head and Sherren.)

months, is a gradual diminution of the area of total analgesia commencing in the proximal parts of the area, and recovery of appreciation of major degrees of temperature, while the area of insensibility to light touch remains unaltered. At this stage, generally completed in about six months, there is still absolute loss of epicritic sensibility, although all the forms of protopathic sensation have recovered. The diffuseness and radiation of pain produced by a pin-prick, or rough handling, or a blow is so unpleasant at this stage that the complete absence of sensation may appear preferable to the patient. The return of sensibility to light touch and the minor degrees of temperature, as well as the power of discriminating between one and two points and of accurately localizing stimuli, rarely commences earlier than six months, no matter how favorable the nerve union may be, and is seldom complete within a year; but even for years careful examination may detect abnormalities.

<sup>1</sup> *Jour. of Physiol.*, 1909, xxxviii, 134.

After incomplete nerve lesions, on the other hand, the appreciation of pain and of light touch return at approximately the same time; it may be taken as an absolute rule that if the simultaneous recovery of protopathic and epicritic sensibility is observed within a few months of the injury the nerve has not been completely divided. Recovery of function is also much more rapid than after complete division of the nerve. It commences at a date which varies with the distance of the injury from the periphery, from about three weeks at the wrist to six months in the plexus, and also with its degree.

Symptoms of *irritation* or perversion of function of sensory fibres are often a prominent feature of disease or injury of the peripheral nerves, especially when the lesion is only partial or when the central stump of a divided nerve is involved in scar tissue. Pain is the most important of these subjective symptoms. Irritation of a nerve by pressure or disease may give rise to severe pain; this is undoubtedly due to the excitation of the sensory fibres by the lesion; the abnormal excitation thus produced is conveyed by the ordinary paths to the higher sensory centres of the cerebral cortex, where it reaches consciousness; its origin is, however, misinterpreted and is referred to the sensory end-organs of the irritated fibres, even although the area in which they lie be completely anesthetic—*anesthesia dolorosa*. Pain of this origin is often paroxysmal, and is generally sharp, burning, or darting; at times, however, it is of a dull aching or boring character; it may be referred to the skin or the deeper structures of the limbs.

*Hyperalgesia* is met with in regions in which epicritic sensation is lost, while protopathic persists, but it often follows partial lesions where there is little or no disturbance of sensation. It not infrequently extends over the boundary of the anatomical distribution of the injured nerve, and is often associated with severe spontaneous pain.

Weir Mitchell gave the name *causalgia* to a condition of severe spontaneous pain associated with hyperalgesia and tenderness, and often with trophic disturbances. It is most frequently seen after bullet wounds; it never results from complete interruption of the continuity of the nerve, and always disappears immediately the nerve is cut across for secondary suture. In these cases it is probable that normal stimuli of peripheral origin are augmented in their passage through the injured region, or it may be that the sensory end-organs become hyperexcitable owing to a disturbance of their connection with their trophic centres. Hyperalgesia appears rarely if ever immediately after the nerve injury, generally not for a period of a few weeks.

Pain is uncommon after nerve injuries by modern high velocity bullets; Olonomakes<sup>1</sup> saw it very rarely accompanying peripheral palsies in the Balkan wars.

Various *paresthesias*, or a feeling of loss of feeling, or of weight or pressure, or, it may be, a sense of warmth or coldness, are frequently due to affection of the peripheral nerves which supply the region to which they are referred; they are generally, but not always, associated with

<sup>1</sup> *Neurolog. Centralblatt*, 1914, xxxiii, 486.

some loss of sensibility. They are, on the whole, met with more frequently in the toxic degenerations of nerves than after traumatic or local lesions; they are probably due to a slight but persistent irritation of the sensory fibres. The most frequent form is generally described as a numbness or deadness of the part, but from the discomfort it causes it is evident that it is more than the consciousness of loss of feeling; it is rather an abnormal positive symptom, which may, even when there is no objective sensory disturbance, seriously interfere with the functions of the part. Even when paresthesias are due to the local injury of a nerve, they are rarely referred to the whole area of its anatomical distribution; in the limbs they tend to be more intense peripheralward.

Occasionally the irritation of one of the terminal branches of a nerve gives rise to pain extending over its whole protopathic distribution, or that of its roots, which may be accompanied by hyperalgesia, or by spasm or contractures of the muscles supplied by it. Neurotomy of the irritated branch gives instant relief.

**Trophic and Vasomotor Symptoms.**—The nutrition and conservation of all tissues of the body are to a certain extent dependent on the integrity of their connection with the central or autonomic nervous systems, and when this is interrupted the isolated tissues may undergo certain changes. The most prominent of these is the atrophy of the muscles which follows the degeneration of the motor nerve fibres which innervate them, but the skin, subcutaneous tissues, and even bone may also undergo structural alterations.

It was for long discussed whether the influence of the nervous system on the tissues is exerted through special trophic fibres, or through the ordinary motor, sensory, or vasomotor nerves. The muscles are certainly dependent only on the integrity of their motor fibres, and the existence of special trophic fibres to other tissues has not been proved. The vasomotor nerves must, however, have a considerable influence on nutrition and growth; parietic vasodilatation follows section or injury of a nerve, and although there may be hyperemia and a slight elevation of the local temperature for a time, the part soon becomes cold, congested, and even cyanosed, owing to the slow circulation through the dilated vessels; and its inactivity leads to lymph stasis.

It seems probable that the trophic centres for the skin and other tissues lie in the spinal ganglia, and that the centrifugal conduction of trophic influence is a function of the sensory fibres; or more probably that the normal nutritional equilibrium is in some way determined by the sensory nerves. The strongest evidence of the trophic function of the spinal ganglia is that the acute changes in the skin which characterize herpes zoster are directly due to disease of them. The innervation of the blood-vessels and of the sweat and sebaceous glands takes place through the sympathetic system; in the limbs, at least, these sympathetic fibres are intimately associated with those of motion and sensation.

*Trophic changes* in the skin vary with the rate of evolution and the degree of the nerve lesion; the more acute it is the more likely are trophic changes to be prominent. They may be met with either in areas of total anesthesia, or associated with only partial disturbance of sensation. In

areas of total analgesia the skin becomes thin and atrophic, and, especially in the hands and feet, inelastic and tightly stretched over the part; owing to the cessation of sweat and sebaceous secretion it is usually very dry and may be either glossy and shiny, or scaly when desquamation of the superficial layers of the epidermis is delayed. In this condition it is very liable to injury, and ulcers often develop, which, as a rule, first appear as blisters or bullæ; these often appear to be the result of the trophic disturbance alone, as they may occur apart from any assignable local cause, or follow a slight bruise. Owing to the loss of sensation the skin is liable to suffer from neglected or unobserved injuries, and to this many of the trophic changes have been attributed. Head<sup>1</sup> has observed that these affections are generally co-extensive with the area of complete analgesia, and disappear with the return of protopathic sensibility.

Another form of cutaneous trophic disturbance which follows incomplete lesions of nerves, and is always associated with spontaneous pain and hyperalgesia (causalgia), was first accurately described by Paget<sup>2</sup> and Weir Mitchell,<sup>3</sup> and by the former termed "glossy skin." It is seen chiefly on the fingers and hands; the skin becomes "smooth, hairless, almost devoid of wrinkles, glossy pink or ruddy, or blotted as if with permanent chilblains" (Paget). The subcuticular tissues shrink, and the skin, which appears tightly drawn over them, is often cracked and the epithelium is partially lost, so that the cutis is exposed in places. According to Weir Mitchell, this condition is often attended with vesicles.

Changes in the growth and condition of the nails often follow nerve injuries. Retardation of growth has been frequently described, but Head has shown that this is independent of sensory disturbance, and that want of movement owing to paralysis or the fixation of the limb is the main factor. Weir Mitchell observed remarkable alteration in the nails associated with glossy skin, which consisted of curving on their long axes, extreme lateral arching, and sometimes a thickening of the cutis beneath their extremities.

When several nerves of a limb are damaged the bones are liable to become fragile, and if it occurs in early life their growth may be retarded. Acute and extensive nerve lesions may lead to swelling of and effusions into joints in the paralyzed regions. Another form of more chronic joint change occurs chiefly in the fingers and wrists; it commences with pain and periarticular thickening, and may ultimately produce ankylosis.

**The Treatment of Local Nerve Lesions.**—This naturally depends on the nature and the degree of the lesion; but there are certain general lines which should be invariably followed. In every case the chief aims should be to maintain the nutrition of the parts, to keep the paralyzed muscles relaxed, and to prevent the occurrence of contracture.

If the nerve has been completely divided, its two ends should be sutured together at once. The prognosis when primary suture is possible is good, but it depends to a certain extent on the nerve injured, the nature of the lesion, the condition of the wound, and the distance of the injury from the periphery; the muscular functions generally

<sup>1</sup> *Brain*, 1905, xxviii, 251.

<sup>2</sup> *Medical Times and Gazette*, 1864, i, 331.

<sup>3</sup> *Gunshot Wounds and other Injuries of Nerves*, Philadelphia, 1864.

return within nine months, but sensation is rarely perfect for two or three years. Often, however, the nerve injury is not observed, or the patient does not come under treatment until after a considerable time, then secondary suture must be performed. The outlook in these cases is not so favorable as after immediate suture, but the time after the injury at which the operation is undertaken, certainly up to three years, seems to have little influence on the recovery (Sherren).

Much depends on the condition of the muscles; when these have entirely lost their galvanic irritability, complete recovery is probably impossible, and according to Warrington and Jones,<sup>1</sup> when the paralysis has lasted several weeks return of full power cannot be expected unless paralyzed muscles have been kept relaxed. It is important that not merely should all scar tissue around the ends of the nerves be removed, but that these should be freshened up and brought into accurate apposition. It is occasionally impossible to bring the divided ends together; then Assaky recommended imposing catgut threads between them to form a scaffold along which the regenerating axis-cylinders may grow, and Vanlair has introduced the method of interposing a tube of decalcified bone or a piece of a small artery or vein for the same purpose. Nerve transplantation, however, seems to give more favorable results in such cases, but, as Merzbacher<sup>2</sup> has shown, the portion of nerve inserted can take an active part in regeneration only when it is taken from the same animal, or an animal of the same species; if taken from another species, it immediately necroses and can consequently at the most act only as a scaffold. In other cases it is preferable to anastomose the peripheral end of the paralyzed nerve into a neighboring healthy trunk, or end to end with a flap raised from the sound nerve. In every case the exact condition of the injured nerve should be examined under an anesthetic if there is complete reaction of degeneration in the muscles it supplies after fourteen days.

Tendon transplantation is occasionally necessary, especially when there is such complete degeneration of the paralyzed muscles that recovery after secondary suture of the nerve cannot be expected. The results obtained are often favorable. It is interesting that after nerve anastomosis and tendon transplantation nerve centres in the cord can acquire new functions or adapt themselves to new conditions.

Whatever surgical treatment is adopted it is important to keep the paralyzed muscles relaxed and prevent shortening and contracture of their antagonists. If the extensors of the wrist and fingers are paralyzed, their joints must be kept fully extended by a splint along the forearm and hand; if it is the flexors of the forearm, the elbow should be held flexed in a sling. But the splint or other apparatus used should be removed frequently, and massage and passive movements carried out.

The next aim should be to maintain the paralyzed muscles in as good a state of nutrition as is possible, and for this electrical treatment is usually employed. If the paralysis is not complete, and if some excitability to the interrupted current remains, faradism may be employed,

<sup>1</sup> *Lancet*, 1906, ii, 1644.

<sup>2</sup> *Neurologisches Centralblatt*, 1905, xxiv, 150.

but it is of little use if the muscles will not react to it, although, according to Mann,<sup>1</sup> regular treatment raises the excitability of both nerve and muscle and increases the flow of blood and lymph. Galvanism is more effective when the muscles will not react to faradism, but the current employed should not be too strong.

Systematic and energetic massage is probably more effective than any form of electrical treatment, and should be combined with it, but the region of the nerve injury should not be forcibly handled. Passive movements are always advisable; and the patient should make efforts to use the paralyzed muscles as soon as any return of power sets in.

### DISEASES OF THE SPINAL NERVES.

**The Cervical Plexus.**—Owing to its deep position among the muscles of the neck, the cervical plexus is rarely injured or affected by disease. The branch which is of most clinical importance is the *phrenic nerve*. Its paralysis may be due to a lesion in the ventral horns of the cord at the level of its origin, to an intraspinal hemorrhage or tumor, or to syringomyelia. Duchenne first described it in progressive muscular atrophy. It is most frequently caused by involvement of the third and fourth cervical roots in meningeal or vertebral disease, especially in spinal caries and syphilitic pachymeningitis. Owing to its deep position the nerve trunk is relatively immune from trauma, but it is occasionally injured by wounds or operations. It has been repeatedly observed after local anesthesia of the brachial plexus.<sup>2</sup> Within the thorax it may be compressed by tumors, aneurisms or enlarged glands. Unilateral paralysis can be often attributed only to a local neuritis, perhaps following exposure to cold. Bilateral palsy sometimes occurs in multiple neuritis, especially in that form which follows the acute infective diseases, in lead poisoning, and in tabes dorsalis (Gerhardt).

The diaphragm is one of the most important of the respiratory muscles; when the patient is at rest its inactivity may give no trouble, but dyspnoea is easily produced by exertion or when respiration is interfered with by disease of the lungs or pleura. If bilateral paralysis sets in suddenly there may be considerable dyspnoea and even cyanosis for a time, but it is quickly relieved by the activity of the accessory muscles. Several cases have been recorded in which accidental injury of one phrenic nerve during operations in the neck has led to a fatal result, but Schröder and Green,<sup>3</sup> who analyzed these cases, come to the conclusion that death was generally due to some other cause.

When there is complete bilateral paralysis the upper part of the abdomen is no longer protruded with each inspiration, but sinks in as the diaphragm is drawn upward by the negative pressure in the thorax; on palpation the descent of the liver and spleen can be no longer felt, and the patient is unable to expand the abdomen by taking a deep breadth. The movements of the thorax are consequently often increased, and the

<sup>1</sup> *Centralblatt f. Nervenheilkunde*, 1897, xx, 1.

<sup>2</sup> *Klauser, Centralbl. f. Chir.*, 1913, xl, 595.

<sup>3</sup> *Am. Jour. Med. Sc.*, 1902, cxxiii, 196.

excessive movement of the lower part of the thoracic cage may draw the abdominal wall tense; this must not be mistaken for the effect of the descent of the diaphragm. If the paralysis is not complete the protrusion of the abdomen in inspiration can be easily resisted by pressure. Owing to inability to take a deep breath there is difficulty in coughing, and the patient cannot spit out with the normal force nor sneeze properly, and there may be difficulty in defecation. Pulmonary symptoms are the most important complication, owing to the relative immobility of the bases of the lungs they may have become dangerously congested.

The symptoms of unilateral paralysis are generally slight and frequently escape observation, but the deficient movement of one side can be definitely determined by a radiograph, or by the direct observation of the movements. The electrical excitability of the phrenic nerves may be examined; they are easily stimulated in the neck between the sternomastoid and scalenus anticus muscles, and above the omohyoid. The phrenic nerves also convey sensory fibres to the pleura, pericardium, and diaphragm, but sensory symptoms have been rarely referred to their disease; in a few cases, however, patients have complained of pain in the mediastinum and in the region of the diaphragm.

The *diagnosis* of diaphragmatic paralysis is not always easy; the examination of the electrical excitability of the phrenic nerves may be very important when other signs are not conclusive. In the majority of the cases the condition is only part of a general peripheral neuritis, or of poliomyelitis. Otherwise, bilateral paralysis is generally due to some disease in the spinal cord or meninges; it can then be rarely an isolated symptom. The vertebral column should be carefully examined, and the existence of tumors in the neck and mediastinum excluded. Paralysis must be distinguished from immobility of the diaphragm owing to diaphragmatic pleurisy or peritonitis, or to large pleural effusions. Or the diaphragm may be weak owing to secondary degenerative changes. Acute fatty degeneration of its muscle fibres frequently occurs in diphtheria, and may enfeeble, but rarely paralyzes the action of the muscle.

**Treatment.**—This should be directed to the removal of the cause if it is possible, but it is rarely so unless it is due to an operable tumor either in the vertebral canal or neck. Otherwise the chief aim should be to avoid all pulmonary complications and save the patient from exertion. If there is reason to suspect a local neuritis, warm fomentations and counter-irritation may be applied over the lower part of the anterior triangle of the neck. Electricity, especially the faradic current, has been employed with apparently some effect; the one electrode should be placed over the phrenic nerve immediately behind the sternomastoid muscles in the neck, the other over the epigastrium.

**Long Thoracic Nerve.**—Isolated paralysis of this nerve is rare, Steinhäusen<sup>1</sup> was able to collect records of only 29 pure cases, it is more frequently found in association with palsy of other muscles of the shoulder-girdle, and it is often seen in progressive muscular atrophy and in the muscular dystrophies. The nerve may be injured in the neck by blows

<sup>1</sup> *Deutsche Zeitschr. f. Nervenheilk.*, 1900, xvi, 399.



or perforating wounds, or by direct pressure from a heavy weight carried on the shoulder; and it occasionally happens that it is bruised by the forcible contraction of the scalenus medius muscle through which it passes, or by excessive stretching of the nerve when the arm is raised above the head, as in painting a ceiling, climbing hand over hand, or hanging suspended by the arms. In the axilla it may be injured by a perforating wound or by operation. Paralysis of the serratus magnus has also been observed after infective disease, and with acute arthritis of the shoulder-joint. In some cases it has been apparently due to a local neuritis following exposure to cold. It is much more frequent in strong muscular men than in women and is more common on the right side. Its paralysis produces very little deformity while the arm is at rest; the scapula may stand slightly higher than normal, with its inferior angle slightly approximated to the vertebral column and separated from the chest wall. When the arm is moved forward into the horizontal position or pressed forward against resistance, the scapula, no longer held against the thorax by the serratus, is rotated on its vertical axis, so that its vertebral border projects backward and appears winged. This deformity is almost pathognomonic of serratus palsy. There is also difficulty in raising the arm above the horizontal, as while the arm is normally abducted from the side as far as the horizontal level by the deltoid alone, its further elevation is brought about by rotation of the scapula, chiefly by the serratus magnus. The latter part of the movement is consequently lost when this muscle is paralyzed, but not invariably, as it can be sometimes carried out by contraction of the middle fibres of the trapezius. There is often diminished muscular power in the whole arm, which disappears when the scapula is firmly bound to the trunk. Slight scoliosis is frequently observed in cases of serratus palsy; it is probably due to an attempt to reestablish the equilibrium upset by the malposition of the shoulder. When the paralysis is due to a neuritis its onset is often accompanied by severe neuralgic pains in the supra-clavicular region, which may radiate up the neck, behind the scapula and even into the arm, but no loss of sensation results from an isolated paralysis of the long thoracic nerve.

**Treatment.**—This must be conducted on the usual lines; it is important to prohibit work or exercise which may produce pressure or strain on the nerve. When the palsy is incurable the humeral attachment of a portion of the pectoralis major may be inserted into the serratus magnus.

**The Suprascapular Nerve.**—Isolated paralysis of it is very rare; Fischler<sup>1</sup> was able to collect only 14 cases. In half of these it was due to direct or indirect trauma, in others apparently to a local neuritis; it may be caused by the pressure of a heavy weight carried on the shoulder, or by a fall on the shoulder or the outstretched arm. Its most prominent symptom is flattening of the infrapinnous fossa and weakness of outward rotation of the humerus owing to the atrophy and palsy of the infrapinnatus; this movement is not, however, completely absent, as the teres minor and the posterior fibres of the deltoid can execute it,

<sup>1</sup> *Neurologisches Centralblatt*, 1906, xxv, 444.

but only very feebly. According to Duchenne, the chief function of the supraspinatus is to act as an elastic ligament in keeping the head of the humerus in close apposition to the glenoid cavity; when it is paralyzed the humerus falls away and the movements of the shoulder-joint are impeded, especially abduction and elevation in the sagittal plane. In every case of its paralysis the patient has complained of weakness and fatigue in the shoulder, and of the inability to carry weights. No definite disturbance of sensation has been detected with suprascapular palsy, but its onset may be accompanied by pain in the shoulder-girdle.

**The Circumflex Nerve.**—Paralysis is most often due to such injury as a blow or fall on the shoulder, dislocation of this joint, or fracture of the upper end of the humerus. Occasionally it is caused by the pressure of a crutch, or by lying for long on the shoulder in deep sleep or in an unconscious state; in those cases in which paralysis follows prolonged operations under anesthesia the lesion is evidently due either to compression or undue stretching of the nerve. Similarly, paralysis of the circumflex has been frequently observed in miners who work lying constantly on the left side. Local neuritis may be due to exposure, or to extension of inflammation from arthritis or from disease in the axilla; neuritis has also been observed in infectious diseases and in diabetes. It is noteworthy that Bernhardt and Buzzard have observed isolated paralysis of the circumflex nerve in lead poisoning.

A complete lesion of the nerve leads to complete paralysis of the deltoid, except of a small number of its anterior fibres, which are supplied by the anterior thoracic nerves. Abduction and elevation of the arm in any plane are consequently impossible, except by rotation of the scapula by the trapezius and serratus magnus. The supraspinatus may also aid in this movement. The shoulder-joint becomes relaxed, and owing to the wasting of the deltoid its shape is altered. The paralysis of the teres minor, which depresses the arm and rotates it outward, is less prominent. The onset is generally accompanied by much pain. Objective sensory disturbances are not constant, but in typical cases the area of anesthesia to light touch occupies an oval area on the outer side of the arm extending from the level of the acromion process rather more than half-way to the elbow; the loss of sensibility to pain and the extremes of temperature is less extensive. When the paralysis lasts for long there is a danger of adhesions forming in the shoulder-joint, and ankylosis may occur. In obstinate cases good results have been obtained by transplanting the clavicular portion of the trapezius and pectoralis major into the deltoid, and by grafting the subscapular nerve into the circumflex.<sup>1</sup>

A little care and the examination of the electrical reaction of the muscle serves to distinguish primary joint disease with the secondary wasting of the muscle from circumflex paralysis.

**The Musculocutaneous Nerve.**—It is rarely paralyzed alone. Bernhardt<sup>2</sup> collected only 14 cases, but as it is not infrequently associated with lesions of other nerves it is important to recognize its symptoms. It is generally due to a blow on, or compression of the arm, or to fracture

<sup>1</sup> Langfeller and Frohse, *Med. Klinik*, 1909, v, 1270.

<sup>2</sup> *Die Erkrankungen der Peripherischen Nerven*, Wien, 1902.

or dislocation of the humerus. When the lesion is complete the biceps and coracobrachialis are absolutely paralyzed, as well as the greater portion of the brachialis anticus. Flexion of the elbow is consequently impossible when the forearm is supinated, but it can be carried out feebly and in limited range by contraction of the supinator longus when the forearm is pronated. Sensory disturbance is limited to the radial side of the forearm and the thenar eminence.

**The Musculospiral or Radial Nerve.**—This is probably paralyzed more frequently than any other nerve in the arm. It may be injured in the axilla by dislocation or fracture of the upper end of the humerus, or involved in callus formation or by the pressure of a crutch. It is occasionally compressed by the head of the humerus when the arm is kept fully abducted and extended during operations under anesthesia (Braun). But it is much more frequently injured during its course round the humerus, very often by the pressure to which it is exposed when a person sleeps on a hard or uneven surface with the arm beneath his body, or with the weight of the head resting on the outer surface of the arm. This occurs so often in a drunken sleep that it has been assumed that chronic alcoholism predisposes to it by lowering the vitality of the nerve (Oppenheim). Gowers has pointed out that it may be injured as it passes through the triceps by a sudden violent contraction of this muscle. Its paralysis has followed the use of an Esmarch's bandage on the arm, and it sometimes results from the injection of ether. A local neuritis due to cold is occasionally assumed to be the cause. When palsy develops during an acute illness it is more probably due to pressure as the patient lies in a semiconscious or delirious state. The affection of some of its fibres is a characteristic feature of lead palsy.

**Symptoms.**—These depend on the site and severity of the injury. When the lesion is in the axilla all the muscles supplied by it are paralyzed and the patient is no longer able to extend the elbow and wrist, or the fingers or thumb at their basal joints, or to supinate the forearm except by the biceps. If the nerve is injured, as it is most frequently, on the outer side of the arm, the extensors of the elbow and more rarely the supinator longus escape, but wrist-drop, which is the characteristic feature of the palsy, is present. The thumb cannot be abducted or extended, but its other movements are intact. Although the flexor muscles are not affected, the hand-grasp is considerably weakened owing to the mechanical disadvantage at which they work when the wrist is not held straight by the normal action of their antagonists; with the hand passively extended its grasp is normal. As the arm hangs by the side, the forearm is generally pronated, and becomes more fully so when the hand grasps any object, owing to the unresisted pronation action of the flexors. The power of supination with the elbow extended is completely lost, but when it is flexed the biceps can supinate the forearm. If the nerve is injured in the forearm, the supinators and even the extensors of the wrist may escape. Paralysis of the supinator longus produces slight weakness of flexion of the elbow.

*Sensory* symptoms are very variable; with the onset there may be subjective sensations of numbness and tingling in its cutaneous distribution,

generally most pronounced on the radial border of the hand. In incomplete lesions, the sensory fibres are, according to the general rule, much less affected than the motor, and even with complete paralysis of the muscle there may be no loss of sensibility; when present the anesthesia is generally most marked over the radial branch. Trophic changes are rarely prominent. In the pressure palsies there may be little or no atrophy of the paralyzed muscles. Occasionally a prominence develops on the dorsum of the hand, which is due either to the swelling of the sheaths of the extensor tendons (Gubler) or to overflexion of the carpus. There may be slight effusion into the carpal joints, and adhesions may form in them. The electrical reactions of the nerve and the muscles it supplies are extremely important; in the pressure palsies the nerve may be inexcitable from above the lesion, while in the portion below it and in the muscles it supplies normal responses can be obtained; the conduction of volitional and electrical impulses is thus interrupted by a lesion which is not of sufficient intensity to damage the continuity or vitality of the nerve fibres. If the lesion is more severe there must be partial or complete reaction of degeneration in the muscles it supplies.

*Paralysis* of this nerve is generally easily recognized, but, as Gowers points out, the fact that it produces loss of extension of the limb at all its joints may lead to error, as palsy limited to a single function suggests central disease; the absence of sensory loss, and of change in the electrical excitability of the muscles may increase the risk of error. In lead palsy it is the musculospiral groups of muscles which are chiefly involved, but the affection is almost invariably bilateral, the onset is usually slow and unconnected with trauma, the supinator longus, as a rule, escapes, and the reaction of degeneration appears early in the paralyzed muscles.

**Treatment.**—This must be conducted on the usual lines. In the pressure palsies the application of the galvanic current for twenty to thirty minutes at a time is of value; the cathodal electrode should be placed over the seat of injury, the anode distal to it, and the strength of current slowly increased until the patient feels it distinctly.

**The Median Nerve.**—Owing to its deep position among the soft tissues of the arm this nerve is much less liable to injury than the musculo-spiral. In the axilla and arm it is usually injured by fractures and dislocations of the humerus and occasionally by the pressure of a crutch, but the most frequent cause of its paralysis is a wound on the palmar surface of the wrist. Occupation palsies frequently involve some or all of the hand muscles supplied by the median nerve; it has been repeatedly observed in laundresses, joiners, milkmaids, and cigarette makers. The exact nature of the lesion is doubtful; often it is undoubtedly due to a neuritis set up by pressure, but in other cases the muscular palsy and atrophy seem to be the direct result of the stress of overwork. Drummer's palsy affects chiefly the thumb muscles, but seems to be generally due to rupture of the tendon of the extensor longus pollicis (Heinicke!).

When the nerve is damaged above the elbow the power of pronating

the forearm is lost; flexion of the wrist is feeble and incomplete, and as it can be performed only by the ulnar flexor, the hand is strongly deviated to the ulnar side. Flexion of the interphalangeal joints is also lost except that of the distal phalanges of the two ulnar fingers, which can be still bent by the unparalyzed part of the flexor profundus. The flexion of the fingers on the metacarpus is unaffected, as it is performed by the interossei. The unopposed extensor action of the latter muscles at the interphalangeal joints may lead to their hyperextension. The thumb is kept extended and adducted by the muscles which remain, and it cannot be opposed nor its distal phalanx flexed; its metacarpal bone comes to lie in the same plane as that of the fingers, like the thumb in the ape. Owing to paralysis of these movements of the fingers and thumb there is considerable difficulty in firmly grasping any object and in employing the hand in any work. When the lesion affects the nerve in the forearm, after its branches to the pronators and flexors have been given off, pronation and flexion of the wrist and fingers may be intact; then it is chiefly the thumb movements which are lost. Bernhardt and Head have pointed out that the branch which supplies the muscles of the hand may leave the main stem of the nerve in the lower part of the forearm and thus escape injury when the wound is at the wrist. When the lesion of the nerve is severe, prominent atrophy of the muscles of the thenar eminence results, and of the flexor surface of the forearm if it is above the elbow. There may be considerable paresthesia in the cutaneous area of the nerve in earlier stages of the paralysis; the occupation palsies and neuritis are generally accompanied by troublesome pain. A complete lesion of the nerve may lead to troublesome trophic disturbance of the skin and nails in the area where protopathic sensibility is lost, and to vasomotor paresis and cessation of sweating in the radial part of the palm. (Fig. 38 *B*, p. 538.)

**The Ulnar Nerve.**—This may be injured alone or with other nerves in the axilla or upper arm by dislocation or fractures of the humerus, or involved in callus formation; occasionally it suffers in crutch palsy. It is much more frequently damaged at the elbow-joint by dislocations or fractures; occasionally ulnar paralysis develops slowly at long periods after elbow injuries owing probably to the pressure of fibrous adhesions or excess of callus on the nerve, and in a few cases it has been due to traumatic or synovial cysts in this region. More rarely the curious condition of dislocation of the ulnar nerve from its groove on the posterior surface of the internal condyle is met with; it probably occurs only when the internal condyle is badly developed. Paralysis due to direct pressure from without is rare; it is occasionally due to pressure on the elbow during sleep, especially in emaciated subjects, or during infective illness. According to Braun, however, these sleep palsies are to be attributed to pressure of the head of the humerus on the nerve in the axilla when the arm is abducted and extended. But undoubtedly wounds in the wrist, which may either injure this nerve alone, or the median nerve and flexor tendons as well, are the most common cause of ulnar palsy. Primary neuritis is rare; a few cases of syphilitic neuritis are described, and leprosy has a predilection for this nerve.

When the nerve is injured at or above the elbow the power of flexion of the hand is very feeble, and when attempted the hand is deviated radialward by the radial flexor; the wrist becomes hyperextended when the fingers are straightened, owing to palsy of its ulnar flexor. The movements of the little finger are lost, the middle and ring fingers cannot be flexed at their distal joints, and owing to the paralysis of the interossei the basal phalanges of all the fingers cannot be flexed or the middle or distal phalanges fully extended. Adduction and abduction of the fingers are also impossible. When the injury is situated in the lower part of the forearm the fibres to the flexor profundus digitorum escape and the interossei and thumb muscles are alone paralyzed; then owing to the unopposed contraction of the long extensors and flexors the hand becomes claw-like—*main en griffe*—with the first phalanges hyperextended and the interphalangeal joints flexed. As the first two lumbricales escape, this deformity is generally not so pronounced as it is in progressive muscular atrophy (Gowers). Adduction of the thumb is also lost. The muscular atrophy which follows severe lesions of this nerve is very typical; the hypothenar eminence disappears, the palm becomes hollow, and the interosseal spaces sink in. When the nerve is severely damaged there is complete loss of sensation in the little finger and over a variable extent of the ulnar border of the hand; on the ring finger and the rest of the ulnar cutaneous distribution sensibility to light touch and the intermediate degrees of temperature are alone affected. As the dorsal cutaneous branch separates from the nerve in the middle third of the forearm it may escape in wounds at the wrist. Dupuytren's contracture of the palmar fascia has been observed after neuritis of the ulnar nerve, and de Leon<sup>1</sup> has described contractures of the ulnar portion of the flexor profundus digitorum, owing to which the three ulnar fingers were bound down in the maximal flexion position.

The chief danger of error in diagnosis is of confusing central and peripheral lesions, as the ulnar nerve contains almost all the root fibres of the eighth cervical and first thoracic segments of the cord.

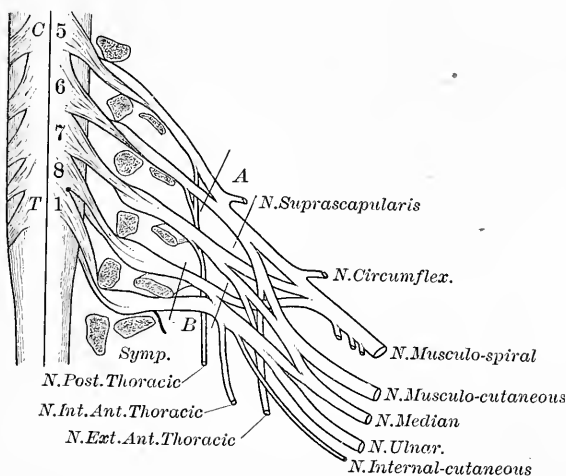
**Paralysis of the Brachial Plexus.**—The brachial plexus is formed by the anterior primary divisions of the lower four cervical and the first thoracic roots; it contains all the fibres which supply the muscles of the shoulder girdles and upper extremities, as well as the sensory fibres to almost the whole of the arm. By division and secondary anastomosis of these roots the three main cords of the plexus are formed. The outer cord receives the ventral trunks of the fifth, sixth, and seventh cervical roots; the inner contains the ventral trunk of the eighth cervical as well as the whole of the first thoracic root, and the posterior cord is made up of the dorsal trunk of the four lower cervical roots. This is the most common form of the brachial plexus, but it is liable to variation, for the fibres concerned in any single function do not constantly leave the cord by the same roots, but although fibres may alter their position relative to the vertebral column, they always maintain their position in relation to other fibres (Herringham). The whole plexus may

<sup>1</sup> *Nouvelle Iconographie de la Salpêtrière*, 1901, xiv, 409.

be shifted up—high or prefixed type—or downward—low or post-fixed type—and the extreme variations between the high and low form of plexus may amount to nearly a whole root (Harris).<sup>1</sup> Paralysis may be due to injury or disease of the nerve fibres in the roots, in the plexus, or in the nerve trunks and branches. Two types of plexus paralysis merit special description.

**Duchenne-Erb Palsy, or the Upper Arm Type.**—In this form the deltoid, biceps, brachialis anticus, and supinator longus are generally paralyzed together, and frequently also the supinator brevis and supra- and infra-spinati; more rarely other muscles of the shoulder-girdle, and the radial extensor of the wrist and the pronator radii teres. The lesion to which it is due involves the fifth and sixth cervical roots either before or immediately after their union, or occasionally the fifth root alone; when

FIG. 39



A diagram of the brachial plexus and its connections with the spinal cord. *A*, represents the usual seat of the lesion in the upper arm (Duchenne-Erb) type of plexus paralysis; *B*, that in the lower arm or Klumpke type. *Symp.* is the Ramus communicans from the first thoracic root which carries the oculo-sympathetic fibres.

caused, as it frequently is, by a blow or fall on the shoulder, it has been assumed that these roots were compressed between the clavicle and the transverse processes of the lower cervical vertebræ, or the first rib, but it seems more probable that the lesion is a rupture of some or all of the fibres of this portion of the plexus owing to excessive stretching and tension. In many cases at least the root fibres themselves are ruptured, or are torn out from the spinal cord. This may be due to any cause which increases the distance between the shoulder and the head and neck. The vulnerability of the two upper roots of the plexus to this form of trauma is explained by the fact that they bend downward immediately on their exit from the spine, so that the extravertebral portion

<sup>1</sup> *Journal of Anatomy and Physiology*, 1904, xxxviii, 399.

of each root forms an angle open downward with the portion which lies in the intervetebral foramen, while in the lower two roots this angle is open upward and in the seventh root there is little or no bend. (Fig. 39.)

In severe cases the arm hangs by the side and cannot be abducted because of the paralysis of the deltoid, or rotated outward at the shoulder if the infraspinatus is affected. Flexion of the elbow is impossible owing to the paralysis of the biceps, brachialis anticus and supinator longus, or it can be effected through a small range by the pronator radii teres and the flexors of the wrists. Supination of the forearm is always weakened by the loss of power in the biceps and it may be impossible if the supinator brevis is also affected. Sensory symptoms are absent in the slighter cases, but there may be paresthesia and loss of sensation on the radial side of the arm and forearm. Even complete section of the anterior primary division of the fifth, and sometimes of the fifth and sixth roots may produce no sensory loss (Sherren). Erb has shown that all the muscles paralyzed in this type of plexus palsy may be made to contract by electrical stimulation over a point in the neck 3 cm. lateral to the sternomastoid and the same distance above the clavicle; the loss of excitability of the nerve trunks from this point is an important sign of the upper arm type of plexus palsy.

**Klumpke Palsy, or the Lower Arm Type.**—This form of paralysis, in which the eighth cervical and first dorsal roots are involved, is generally met with only as the residue of a more extensive lesion, but it may be due to compression by a tumor in the neck or a growth in connection with the lung or vertebral column. Owing to the deeper position of these roots they are rarely injured by trauma. The palsy is characterized by an atrophic paralysis of the intrinsic muscles of the hand and generally of some of the forearm muscles, especially the flexors, with a certain amount of loss of sensation on the ulnar fingers and the ulnar border of the hand and forearm. Frequently, too, there are pupillary symptoms due to injury of the sympathetic fibres which leave the cord in the first thoracic root; but as these branch off from the root immediately outside the intervertebral foramen, they are affected only when the inner portion of the root is damaged. When they are injured, the palpebral fissure is narrowed, the pupil contracted, and there may be a slight degree of exophthalmos on the same side as the arm palsy. Vasomotor paresis on the same side of the face occurs probably only when the second and third thoracic roots are injured (Klumpke). When the small hand muscles are alone paralyzed, the typical deformity of claw-like hand results; if the flexors of the fingers and wrist are also powerless, the wrist may be hyperextended owing to contracture of the extensors.

These traumatic plexus palsies are frequently followed by severe spontaneous pain in the arm, probably owing to the constriction of the nerve fibres in scar tissue, and often by trophic disturbances.

**Birth Palsies.**—Birth palsies due to injury of the brachial plexus generally belong to the Duchenne-Erb or upper arm type, and the limb cannot be adducted or rotated outward at the shoulder, or flexed or supinated at the elbow. The paralysis is often more extensive, and occasionally it is one or more of the lower roots which is damaged. The



causation of these birth palsies has been much discussed; for the purpose of treatment it is important to recognize the nature of the lesions and to what they are due. Fracture or separation of the epiphysis of the humerus, dislocation of the shoulder, and fracture of the clavicle are occasionally found associated with birth palsies, but they are rarely the direct cause. In some cases injury of the plexus may be due to the direct pressure of the accoucheur's finger in the axilla or supraclavicular fossa, or to the blade of the forceps in the latter region; or the roots are compressed between the clavicle, which is drawn backward by the elevation and retraction of the shoulder, and the transverse processes of the lower cervical vertebræ or the first rib. It is most probable that the majority of these birth palsies are due to laceration or rupture of the fibres of the nerve trunks or roots by excessive stretching and tension or by the forcible deviation of the head and neck away from the shoulder which is temporarily fixed (Fairbank<sup>1</sup>). Clarke, Prout, and Taylor<sup>2</sup> found rupture of the sheath and hemorrhages into the nerve, with secondary cicatricial contraction, in cases which were operated upon; complete rupture of the fifth root alone has been observed, but in most cases the lesion is found at the junction of the fifth and sixth roots. The sensory disturbances are slight and generally unimportant, but marked trophic changes may result. The paralyzed muscles atrophy quickly and lose their electrical excitability.

*Prognosis.*—The outlook in the plexus palsies naturally depends very largely on the nature of the lesion, but it is much less favorable than in disease of the peripheral nerves. Only six of the twenty-three cases recorded by Bruns recovered, and eleven of the thirty reported by Warrington and Jones.<sup>3</sup> On the other hand Fairbank believes the majority of birth palsies recover. Many of the cases of neuritis, but these are rare, and of those in which the loss of conductivity of the fibres is due to pressure or bruising, improve or make complete recoveries; but when a root or trunk is torn or ruptured the prognosis is very grave unless surgical treatment can be successfully adopted. The ruptured fibres become enveloped in cicatricial tissue, and the cells of origin of the fibres in the ventral horns of the cord are liable to degenerate completely when their axis-cylinders are forcibly torn out. The prognosis is consequently less favorable in cases of rupture due to indirect violence than when the plexus is damaged by a penetrating wound, and the chances of recovery diminish the nearer the lesion is to the spinal cord.

*Brachial Neuritis.*—This is a form of neuritis limited to the brachial plexus which is closely analogous to sciatica; it seems relatively rare. It occurs generally in late life and in females more often than in males. The pain, which is often sudden in onset, is generally of great severity and is at first referred to the back of the scapula, the forearm or hand, or to the region of the plexus itself above the clavicle or in the axilla. As a rule, it is at first intermittent, but soon becomes more continuous with proxysmal variations, which occur spontaneously or are induced by movement. When fully developed the pain may spread over the

<sup>1</sup> *Lancet*, 1913, i.

<sup>2</sup> *Am. Jour. Med. Sc.*, 1905, cxxx, 670.

<sup>3</sup> *Lancet*, 1906, ii, 1644.

whole arm; it is usually dull and aching, but in the severer attacks it is a sharp, lancinating or stabbing. It may be accompanied by undue sensitiveness or hyperalgesia of the skin, and perhaps with tingling and numbness, but definite objective change in sensation is rare. The muscles may present slight wasting and loss of power, but it is more apparent than real, as the patient is reluctant to move the limb owing to the pain which ensues. Adhesions often develop in the joints and trophic changes in the skin and subcutaneous tissues. The condition can be mistaken for neuralgia; the chief points of distinction are the persistent tenderness of the nerves, and the influence of movement on the pain. There seems to be occasionally a danger of mistaking brachial neuritis for pain of angina pectoris or that due to an aneurism, and especially for arthritic changes in the shoulder-joint. The chief aim in treatment should be to avoid movement or anything which may excite pain. The disease is obstinate, and complete recovery cannot be always expected.

*Treatment.*—In neuritis counter-irritation may be applied over the plexus. It is much more important to maintain the nutrition of the paralyzed muscles by electrical (galvanic) treatment and massage, and to prevent the formation of adhesions in the joints and of contracture in the muscles. Warrington has laid emphasis on the importance of preventing the overstretching of the paralyzed muscles by the unrestrained contraction of their antagonists, and Fairbank recommends strongly the use of a splint by which the arm may be held abducted from the side and elevated. When there is evidence that the nerves have been ruptured, or that the damaged fibres are enveloped in cicatricial tissue operative interference becomes necessary. If there is no return of power within nine or twelve months of the injury in a case which has had proper treatment, it is justifiable and generally advisable to explore the plexus. The results of surgical treatment have been less favorable in cases of birth palsy than in the traumatic palsies of the adult, but a considerable number of cases of both varieties have been reported in which more or less complete recovery has followed secondary suture or grafting of the nerves. Winnen<sup>1</sup> found that recovery or some improvement occurs in 70 per cent. of the operated cases.

**Symptoms Due to Cervical Ribs.**—Supernumerary cervical ribs are not rare; they are generally bilateral but in the majority of the cases the symptoms they produce are limited to one side. It has been generally assumed that these are due either to abnormal stretching, or to compression of the lower roots or the lower cord of the plexus as these pass over the rib, but Sargent<sup>2</sup> believes that the nerves are in closer relationship to the fibrous band which connects the free end of the rib with the sternum or with the first thoracic rib, and that they are damaged by this as the rib rotates in the respiratory movements. Movements of the arm may also increase the compression of the nerves by this band. Many persons with cervical ribs never develop any characteristic symptoms, probably due to the fact to which Eisler has drawn attention, that cervical ribs are generally associated with a plexus of the prefixed type

<sup>1</sup> *Deutsche Zeitschr. f. Chir.*, 1911, cxxiii, 903.

<sup>2</sup> *Proc. Roy. Soc. of Med.*, 1913, vi, Clin. Sect., 117.

which receives few or no fibres from the first dorsal root. On the other hand cases with the characteristic symptoms of cervical ribs occur in which no supernumerary ribs exist, and in which the clinical disturbances are due to the pressure of a normal or underdeveloped first thoracic rib on a postfixed plexus. The determining factor is consequently a disturbance in the relative development of the rib elements and the formation of the plexus (Wood Jones<sup>1</sup>).

The vascular symptoms are also generally assumed to be due to compression, elevation and stretching of the subclavian artery by the rib, but Todd<sup>2</sup> believes they must be attributed to compression of the sympathetic vasomotor fibres in the lowest cord of the plexus.

The shape and direction of growth of the rib are of more importance than its size; those that grow directly outward are not so liable to cause symptoms as those that project downward and forward into the posterior triangle of the neck, and which may consequently seem relatively insignificant on *x*-ray examination.

FIG. 40



To show wasting of the thenar muscles in a case of cervical rib.

The symptoms are much more common in females and generally appear first in early adult life though the ribs exist from childhood; this late development depends on the fact that as growth proceeds the shoulders drop, and more in females than in males, and that consequently the tension on the affected roots increases. The more energetic use of the upper limbs in adult life is also an important factor. The more frequent occurrence of the symptoms in the right arm than in the left, and the considerable relief that is often obtained by raising the shoulder, are to be explained by this factor.

The first symptom, and the only one in many cases, is almost invariably *pain* in the arm, which is generally located in the ulnar border of the forearm, hand and fingers, but is occasionally referred to the radial aspect of the limb or to all the fingers; it is always increased by vigorous or continued movement of the limb. It may be associated with numbness and tingling, or a feeling of coldness in the same areas, usually most pronounced in cold weather. In the majority of the cases some loss of

<sup>1</sup> *Proc. Roy. Soc. of Med.*, 1913, vi, Clin. Sect., p. 97.

<sup>2</sup> *Jour. Anat. and Physiol.*, 1911, xlvii, 250.

sensation eventually appears in the distribution area of the first thoracic root, or on the areas of the first thoracic and eighth cervical roots, that is on the ulnar border of the forearm and hand, and in the two ulnar fingers. As is characteristic of root lesions, there is generally a certain amount of dissociation of the various forms of sensation, the tactile loss being less pronounced than the analgesia, while the thermal sense seems to suffer most severely. In many cases there is little or no objective sensory disturbance.

A feeling of weakness or uselessness of the limb is often one of the earliest symptoms, but definite motor paralysis is much less common than the sensory disturbances. After a variable period, during which the pain has been more or less constant, the hand generally becomes weak and its intrinsic muscles, especially those of the thenar eminence, atrophy and lose their normal electrical reactions. In other cases there is a general wasting of the small hand muscles, and the hand may thus assume the *main en griffe* position. The flexors of the fingers, and to a less extent those of the wrist, may be also affected. Spasm of the flexors of the fingers and probably of other muscles frequently occurs when the palsy is not advanced. Clonic twitching has been observed.

The almost constant absence of oculo-pupillary symptoms, despite the paralysis of the first thoracic nerve, indicates that the lesion is situated outside the intervertebral canal where the sympathetic fibres branch off from the root. Sympathetic ocular symptoms have, however, been observed. The vascular symptoms, which are rarer, are inequality of the radial pulses, cyanosis and coldness of the limb, but arterial thrombosis and even gangrene of the fingers have been observed. The cervical rib may be often palpated in the neck, and there is generally a point of tenderness to pressure over it. Cervical scoliosis convex toward the rib was present in 22 of 61 cases collected by Schönebeck.

The *diagnosis* must depend largely on a radiographic examination, by which the presence of abnormal ribs may be easily revealed; but it must be remembered that cervical ribs often produce no symptoms and that other nervous diseases are occasionally associated with them, especially syringomyelia, which has been found in several cases. Cervical ribs should be suspected in every case of isolated paralysis of the first thoracic, or of paralysis of the first thoracic and eighth cervical, roots, which cannot be otherwise explained. The presence of vascular symptoms, as inequality of the radial pulses, is of considerable value.

The only rational *treatment* is the removal of the supernumerary rib; the operation is attended with little danger, though more extensive paralysis has resulted from operative interference with the plexus. Of 87 cases collected by Streissler<sup>1</sup> 77 per cent. recovered and an additional 13 per cent. improved. The pain is almost invariably relieved by removal of the rib, but it may disappear if the limb is supported or kept at rest. The recovery of the muscles is slower and frequently incomplete.

**Nerves of the Lower Limbs.**—These are much less frequently involved in injuries or affected by disease than those of the upper extremities.

<sup>1</sup> *Ergeb. d. Chirurg. u. Orthoped.*, 1913; v, 280.

**The Anterior Crural Nerve.**—Isolated palsy of this nerve is rare, but it may be due to compression by abdominal growths or by a psoas abscess, or to injury by fractures of the upper end of the femur or of the pelvis, or disease of these bones. Primary local neuritis is seldom seen, but occurs occasionally in diabetes. This nerve, either alone or with the obturator, may be injured during parturition; this probably occurs with greater frequency than is recognized, owing to the rapidity with which it recovers from slight compression and to the fact that its symptoms may not be noticed while the patient is confined to bed. Ernst has observed 30 cases in 800 births. The psoas muscle is paralyzed only when the nerve is damaged in the immediate neighborhood of the lumbar plexus; when the lesion is situated here the thigh cannot be flexed on the abdomen, and if paralysis is bilateral the trunk cannot be flexed on the thighs when these are fixed; the patient is consequently unable to rise from the supine position. When the lesion is in the intra-abdominal portion of the nerve the iliacus alone is paralyzed, and flexion of the hip is only weak. The most prominent symptom is paralysis of the extensors of the knee, and the absence of the knee-jerk in the affected limb. Paralysis of these muscles does not make standing or walking impossible, but contraction of its flexors must be avoided, as the patient cannot resist flexion or actively straighten the joint. The paralysis of the pectineus and sartorius does not produce any other prominent symptom.

Its sensory branches arise in the upper part of the thigh; the middle and internal cutaneous are distributed to the lower two-thirds of the front and inner side of the thigh, the internal saphenous to the front and inner side of leg, and the inner side of the dorsum of the foot. Disturbance of sensation, paresthesia, or radiating pains may be present over these areas when the nerve is injured at or above the level of the groin.

The *diagnosis* is, as a rule, evident; the only risk is of confusing the marked atrophy of the quadriceps extensor group on the front of the thigh which occasionally results from inflammation of the knee-joint with atrophy of these muscles, due to a nerve lesion; but in the former, although the electrical excitability of the muscles may be diminished, there is no reaction of degeneration. The ordinary lines of *treatment* must be followed; good results have been obtained by grafting the tendon of one of the flexors of the knee into the tendons of the quadriceps extensor when the paralysis is permanent.

**The Obturator Nerve.**—This is still more rarely injured alone. It may be injured during parturition, by intra-abdominal or pelvic growths, or by an obturator hernia. When the muscles it supplies are completely paralyzed the limb cannot be adducted, and although it can be raised by flexion of the hip, it cannot be thrown across its fellow when the patient is seated on a chair. Owing to paralysis of the obturator externus, outward rotation is enfeebled, and inward rotation of the thigh is also weak, as the adductor magnus is paralyzed. Gait is not seriously interfered with. The disturbance of sensation is limited to a small area on the inner side of the lower half of the thigh.

*Meralgia paresthetica* is a condition characterized by paresthesia and pain, usually with slight objective disturbance of sensation, in the region

supplied by the external cutaneous nerve on the front and outer side of the thigh. It occurs chiefly in middle-aged men, less frequently in women. Its etiology is obscure; in many cases there has been a history of trauma; probably the long course of the nerve through the fascia predisposes to its injury. In one case a localized perineuritis was found but in other specimens there was no abnormality. In about one-sixth of the cases reported the condition was bilateral. In 1900 Schlesinger<sup>1</sup> analyzed 122 recorded cases, and in the same year Musser and Sailer<sup>2</sup> added ten personal observations in a valuable contribution. The condition has been associated with flat foot. The symptoms vary greatly in intensity but the most common complaint is of abnormal sensations, of numbness, coldness, or tingling, on the front and outer surface of the thigh. In other cases pain is the chief symptom; it may be very severe, but is usually felt only after walking or standing; it is probably due to constriction of the nerve as it passes through the deep fascia when the latter is tense. Occasionally it persists even when the patient is lying down. In the majority of the cases there is a point of tenderness just below the anterior superior iliac spine where the nerve pierces the fascia. Objective sensory disturbances are very variable, but often considerable; occasionally there is hyperesthesia. The symptoms are very intractable; rest may be necessary when there is much pain, and massage and the faradic brush over the course of the nerve may give relief. Resection of the nerve has cured some cases, but in others the pain has returned (Bramwell).

Similar symptoms have been occasionally observed in the distribution of the middle cutaneous branch of the anterior crural nerve, but generally associated with meralgia paresthetica. Lasarew,<sup>3</sup> who has found the condition isolated, has given it the name *meralgia paresthetica anterior*.

**Gluteal Nerves.**—Isolated paralysis of the *superior gluteal nerve* is uncommon; the muscles it supplies are the chief abductors and inward rotators of the thigh, and when they are paralyzed these movements are weak or lost. As the posterior fibres of the glutei rotate the limb outward, this movement becomes weak. The *inferior gluteal nerve* is rarely paralyzed alone; when this occurs the thigh cannot be forcibly extended, nor the trunk straightened on the thigh when the lower limbs are fixed. Standing and walking on a level are not seriously interfered with, but the limb is of little use in ascending steps, and the patient has difficulty in rising from the sitting position.

The **sciatic nerve** supplies motor fibres to the ham-strings and to all the muscles below the knee, as well as the skin in the outer side of the leg and the whole of the foot except a small part of the inner portion of its dorsum. Its main terminal branches are the external popliteal or anterior tibial nerve, and the internal popliteal or posterior tibial nerve; these generally separate in the popliteal space, but are sometimes distinct from their origin in the plexus.

<sup>1</sup> *Centralbl. f. d. Grenzgeb. d. Med. u. Chir.*, 1900, iii, 241.

<sup>2</sup> *Journal Nerv. and Ment. Dis.*, 1900, xxvii, 16.

<sup>3</sup> *Deutsch. Zeit. f. Nerven.*, 1908, xxxiv, 154.

**The External Popliteal Nerve** may be injured by direct trauma in any part of its course, but it is especially liable to be bruised by a blow or by pressure as it bends round the fibula. It has been occasionally torn or ruptured by violent extension of the limb, and paralysis has been frequently observed in laborers who work in a kneeling or crouching position; there it is probably due to compression of the nerve between the fibula and the tense tendon of the biceps cruris. A primary neuritis of this nerve is by no means rare, and even in a general neuritis its fibres seem especially liable to degeneration. Lead palsy is occasionally limited to its distribution, especially in children (Putnam), but in these cases the tibialis anticus escapes as a rule. Isolated paralysis of this nerve is sometimes seen in *tabes dorsalis*.

The whole limb must be unduly raised, as it is brought forward in walking, to enable the toes to clear the ground. Inversion and eversion of the foot are also weakened when there is a total paralysis. When the tibialis anticus is alone paralyzed the foot can be still flexed by the long extensors of the toes, but it is at the same time abducted; while if the function of the latter muscle alone is lost the active tibialis anticus inverts and adducts the foot. As Bernhardt points out, it not infrequently happens that the tibialis anticus escapes when the other muscles of this group are paralyzed. The paralysis of the peroneus longus is most evident when the ankle is extended, as the foot is then so strongly inverted by the unopposed action of its extensors that its outer border rests on the ground, and, as the inner part of the foot is not supported during extension, flat foot may develop. When all these muscles are paralyzed talipes equinus gradually develops, owing to contracture of their antagonists; if the paralysis of all the muscles is not equal in degree the foot may be at the same time either inverted or everted, according to the degree of the paralysis of the muscles with the opposing function. The toes may be permanently flexed by the contracture of the unopposed flexors and interossei. The sensory loss which results from a lesion of this nerve is limited to the outer side of the leg, the dorsal surface of the foot, and the dorsum of the first phalanges of the toes.

**The Posterior Tibial or Internal Popliteal Nerve.**—Owing to its deeper course it is less liable to injury than the external popliteal, and its isolated paralysis is consequently much rarer. A few cases have been recorded in which it has been injured by the tendons of the flexors of the knee when these muscles are forcibly contracted (Oppenheim), and it may be compressed by or involved in tumors, aneurisms, or inflammations.

The most prominent feature is inability to extend the foot or flex the toes, so that the patient can no longer stand on tip-toe or spring from the forepart of the foot in walking; if the paralysis is of long duration the unantagonized action of the flexors of the ankle produces talipes calcaneus, while the unopposed action of the peroneus longus leads to eversion of the foot and increases the plantar arch. Flexion of the distal and middle phalanges of the toes is no longer possible, owing to paralysis of the long flexors, while the loss of the interossei and of the adductors and abductors of the great and small toes makes lateral movements

impossible. The unopposed contraction of the long extensors may lead to permanent overextension of the basal phalanges.

When the conduction of sensory impressions is completely interrupted there is loss of sensation on the outer side and back of the lower third of the leg, on the outer border of the foot and on the sole and plantar surfaces of the toes, as well as in the dorsum of the distal phalanges. There may be trophic disturbances and ulcers may form.

**Paralysis of the Main Trunk of the Sciatic Nerve** may be produced by fractures of the pelvis or of the upper end of the femur, or by dislocations of the hip-joint; or the nerve may be compressed by tumors in the pelvis or invaded in the extension of septic processes from the surrounding tissues. Some or all of the fibres may be paralyzed during parturition, but the lesion is then generally situated in the lumbosacral plexus; the sciatic nerve of the child may be injured by traction on the leg in breech presentations. The symptoms of complete paralysis by a lesion near the sciatic notch are those of paralysis of its terminal branches, the internal and external popliteal nerves, with, in addition, palsy of the flexors of the knee. When the latter are powerless the limb must be held extended at the knee in walking, and it can be used only as a stilt; there is no power of movement at the ankle-joint. As all the sensory fibres enter its terminal branches, the loss of sensation in a complete sciatic palsy includes the outer side of the leg and the whole of the foot except a small area in the inner side of its dorsum.

**The Lumbar and Sacral Plexuses.**—Isolated paralysis of the lumbar plexus is extremely rare and merits no further reference, but palsy of the whole or part of the sacral plexus is occasionally met with. It may be due to invasion or compression of some or all of its roots by tumors or inflammation, or the roots may be injured by pressure from the fetal head during birth. In the latter case it is generally only the fibres which enter the external popliteal nerve which suffer. This, it has been shown by Hünemann<sup>1</sup> and Thomas,<sup>2</sup> is due to the fact that the higher roots of the plexus, from which this branch receives the majority of its fibres, lie directly on the bone as they pass over the brim of the pelvis and are consequently more liable to suffer from compression than the sacral roots which are separated from the bone by the pyriformis muscle. For the same reason the superior gluteal nerve is often injured at the same time. But probably the most common causes of paralysis of these roots are malignant tumors of the pelvis, or tuberculous caries, sarcomata or metastatic carcinomata of the sacrum, which either compress or invade these roots in the intervertebral canals. The symptoms are generally those of an incomplete sciatic paralysis, but if the upper roots are involved the outward rotators of the hip and the gluteal muscles are in addition paralyzed; or if the lower, there will be probably sensory loss in the distribution of the small sciatic nerve on the back of the thigh and on the buttocks and perineum.

**Diagnosis.**—The diagnosis of disease of the nerves of the lower limbs is, as a rule, easy, but different conditions with which a partial or complete

<sup>1</sup> *Archiv f. Gyn.*, 1892, xlii, 489.

<sup>2</sup> *Johns Hopkins Hosp. Bull.*, 1900, xi, 279.



paralysis of the sciatic nerve and its branches may be confused needs further consideration. The diagnosis is often greatly dependent on the history of the mode of onset of the paralysis and of its course; when it immediately follows an injury in the region of the nerve there can be little room for doubt, if the symptoms correspond to the portion of the nerve injured. Disease of the sciatic nerve and its branches must be distinguished from:

1. *Lesions of the Sacral Plexus and of the Extradural Portions of the Lumbosacral Roots.*—When the disease is situated in the sacral plexus, muscles other than those supplied by the sciatic nerve are paralyzed, as the glutei, the obturator internus, the gemelli, and the quadratus femoris; and the anesthesia may extend to the back of the thigh and to the buttocks if the lower portion of the plexus is involved. A careful examination of the pelvis may reveal the presence of a tumor or of other disease. The extradural portions of the sacral roots are most frequently involved by tumors or disease of the sacrum; at first, as a rule, only one root is affected and the earliest symptom is generally pain, which is often extremely severe, referred to the peripheral distribution of its sensory fibres, and paresis of the muscles supplied by it. The neighboring roots are subsequently paralyzed, and if the disease extends across the middle line, motor and sensory symptoms may develop in the opposite limb. The distinguishing feature, as contrasted with a plexus or nerve paralysis, is that the motor and sensory symptoms correspond in extent with the distribution of the root fibres. Further, if the lower sacral roots are involved before they give off their visceral branches the bladder and rectum are paralyzed; true sphincter paralysis never results from disease of the nerves.

2. *From lesions of the cauda equina*, the paralysis of the nerves which spring from the sacral plexus may be distinguished by the fact that the symptoms in the former are always of radicular and not of nerve distribution, that they are almost invariably bilateral, and that when the disease has advanced sufficiently far all the roots below the level of the intrathecal disease are generally involved. The sphincter functions too are almost invariably affected. It is more difficult to distinguish between disease of the cauda equina and of the extradural portions of the spinal roots; in the latter condition, however, the symptoms are often uniradicular for a considerable time, as the sacral disease to which they are most commonly due will generally involve only one root at first. Another point of distinction is that all the roots below the level of the affected one are not paralyzed in the latter condition, no matter how long the disease lasts, unless the sacral tumor extends into the vertebral canal and compresses the cauda equina; while when this is primarily affected by tumor or meningitis, all the roots which pass through the level of the disease are, as a rule, compressed.

3. *From disease of the sacral segments of the cord* the diagnosis is easier. The symptoms, as a rule, develop more rapidly; they are almost invariably bilateral and are typically segmental in distribution, but all functions represented in the segments below the upper level of the disease are interfered with. If, however, the lower segments are not involved,

the paralysis of the muscles which they supply is not associated with atrophy or change in the electrical reactions. The severe radiating pains which are an almost invariable symptom of root lesions are absent, and anesthesia develops earlier. The sphincter functions are generally seriously affected.

**Sciatica.**—This term is commonly applied to all affections of which the chief symptom is pain in the distribution of the sciatic nerve. Such pain may be of the nature of a neuralgia and unassociated with any disease of the nerve, or it may be due to a neuritis, or to compression of the nerve or its roots by tumors or by fibrous adhesions secondary to inflammation. It is unfortunate that the one term should be used for the symptoms of these different conditions, but it is, indeed, often difficult to differentiate between them. It is, however, important to separate the cases in which there is pain without any evidence of organic disease in the nerve, from those in which sciatic pain is associated with symptoms of a nerve lesion, as anesthesia, atrophic muscular paresis, change in the electrical reactions and loss of the Achilles tendon-jerk.

**Etiology.**—Males are affected much more frequently than females, in about the proportion of 5 to 1. It occurs more frequently in middle life, and very rarely, if ever, under fifteen years of age. It has been attributed to almost innumerable causes, but exposure to wet and cold is generally the only apparent exciting factor; it may follow sleeping in a damp bed, or sitting on a wet or cold seat. Gowers insists that many cases develop on a gouty diathesis; others undoubtedly follow spondylitis. The disease may be also due to trauma to the nerve, as by continuous pressure on the edge of a chair, a fall on the buttock, or injury in the neighborhood of the hip-joint. Occasionally an attack sets in after severe muscular exertion, but probably only in those predisposed to the disease. It occurs frequently in anemic and badly nourished subjects, and in the course of chronic intoxications (alcohol), without any apparent exciting cause; and often after infectious diseases. Sciatica may be a symptom of diabetes, and is then usually bilateral. Quénu has shown that the pain may be due to the pressure of varicose veins on the nerve in the neighborhood of the sacrosciatic foramen; this generally occurs only in those who work all day standing erect.

Sciatic pain may be also due to the presence of tumors or inflammatory processes in the pelvis, or to a loaded rectum, which may either directly compress the nerve or affect the nutrition by the venous stasis it produces. Finally, pain in the course of the sciatic nerve may be due to affection of its roots by disease of the sacrum or lesions of the cauda equina. Hysterical sciatica has been described.

**Symptoms.**—The chief symptom is pain along the course of the nerve, or limited to one of its chief branches. The onset is occasionally sudden and associated with slight pyrexia and constitutional disturbances, but, as a rule, it sets in gradually with pain in the buttock or back of the thigh in movements or in postures which make the nerve tense or cause pressure upon it. In other cases the onset of the typical severe pain is preceded by slighter diffuse pain or a feeling of discomfort during walking or after exercise. The pain increases gradually in severity;

it may be either gnawing and burning, or sharp and darting in character. As a rule, it is constant, but severer paroxysms occur, either spontaneously or excited by movement of the affected limb, and its intensity generally increases at night. It may be at first limited to one portion of the nerve, generally that in the upper portion of the thigh, but as the disease develops it usually extends along the whole length of the sciatic trunk and its branches. Often bouts of pain occur which shoot from the buttock down the limb; such attacks may be described by the patients in similar terms to the lancinating pains of *tabes dorsalis*. It is usually most intense in certain points, as over the sciatic notch, in the middle of the thigh, in the popliteal region, below the head of fibula, and behind the external malleolus; more rarely it is referred to the region of the postero-superior iliac crest, or is most severe in the foot. The seat of the chief pain is often, however, variable in any case from day to day. It is generally more or less accurately limited to the course of the nerve, but in other cases it is referred to its whole cutaneous distribution. The most comfortable posture is lying on the back or on the affected side, with the thigh slightly flexed and the knee considerably bent, and when sitting the patient generally rests only on the tuber ischii of the unaffected side with the hip-joint of the painful limb extended as much as possible. In walking the hip and knee are held in moderate flexion with the foot extended at the ankle-joint, and only its toes and forepart touching the ground. Any sudden movement may bring on an attack of pain.

The disease is further characterized by the extreme tenderness of the nerve to pressure, rarely absent, except, according to Edinger, in those cases in which the sciatica is due to compression of the nerve by distended veins, and in which the spontaneous pain disappears when the patient lies at rest. In some cases the muscles of the limb are also tender to pressure. It is characteristic of the pain that it can be invariably produced by stretching the nerve; this can be most easily done by flexing the thigh with the knee extended, or by extending the knee when the hip is flexed—*Lasègue's sign*, or, as Gowers has shown, by pressure on the nerve in the popliteal space as the patient sits in a chair with the knee flexed to a right angle; the pain which is produced by either of these means is felt not only at the point of pressure, but along the course of the nerve in the back of the thigh. Bechterew has pointed out that full flexion of the opposite hip when the limb is extended at the knee also often produces pain, but believes this indicates disease of the roots or of the cauda equina.

The pain is usually associated with *paresthesia*. The muscles supplied by the sciatic nerve and its branches often become flabby and undergo a slight degree of general wasting, even in cases which are not due to any organic lesion, when the disease is of long duration; but in this class of cases there is no change in the electrical excitability of the muscles. Severe reflex spasms of the limb and cramps in some of its muscles, especially in the calves, are not infrequently observed.

Slight trophic and vasomotor disturbances are occasionally met with, but usually only pallor, dryness, and coldness of the skin. In other cases there may be an increase of the surface temperature, and of sweat

secretion. Herpes has been observed. Scoliosis of the lumbar spine convex to the affected side is often seen, but its immediate cause is in dispute; it may be due to an attempt to spare the painful limb by tilting the centre of gravity toward the opposite side, or to a relaxation of the lumbosacral muscles of the affected side. All explanations meet with the difficulty that the scoliosis is occasionally in the opposite direction, concave to the side affected. Kyphosis also occurs, but only rarely.

In addition to the pain and tenderness, evidence of organic disease is present in a considerable proportion of the cases. There may be diminution of cutaneous sensibility, but, as a rule, it is nothing more than a blunting of tactile sensation on the back of the leg and on the foot. When there is an organic nerve lesion the hamstrings and leg muscles and occasionally the glutei may be found not merely flabby, but distinctly wasted and weak; but the feebleness of movement due to a true paresis must be distinguished from the reluctance of the patient to exert full power owing to the fear of pain. The most certain indication is the presence of qualitative changes in the electrical reactions of the wasted muscles, which must be regarded as proof of the existence of an organic lesion of the nerve. The third sign of the existence of organic disease in the nerve is the absence of the Achilles tendon-jerk. It seems very doubtful if this reflex ever disappears in the purely neuralgic cases; in fact, in many such cases it is very brisk or even exaggerated; as both the afferent and efferent paths of the reflex arc are contained in the sciatic nerve, its diminution or disappearance is one of the most delicate signs we possess of the presence of organic disease of the nerve; probably this may be excluded in all cases in which the reflex is undiminished. The reflex seems to be absent in about 30 to 40 per cent. of the cases which are clinically regarded as sciatica (Strasburger<sup>1</sup>).

In the great majority of the cases sciatica is unilateral, but it is occasionally bilateral, in about 7 per cent. of all cases according to Gibson's<sup>2</sup> statistics, though Hyde<sup>3</sup> found it in 33 per cent. of his cases. This, as a rule, indicates a general and not a local exciting cause; it occurs frequently in diabetes, and may be a part of an incomplete general neuritis.

Sciatica has been regarded by some recent authors as a disease of the dorsal roots of the cauda equina. Dubois<sup>4</sup> pointed out that the hypoaesthesia, when there is any, may correspond in extent to the distribution of one or more of the sacral roots, and Lortat Jacob and Sabaréanu<sup>5</sup> confirmed his observations. The sensory loss, according to these observers, most commonly coincides with the cutaneous areas of the last lumbar and the upper two sacral roots. In at least two of their six cases, however, there was an earlier syphilitic infection, and in one of these a lymphocytosis of the cerebrospinal fluid made probable the existence of a syphilitic meningitis. The fact that the small sciatic nerve, and more rarely the anterior crural and the perineal nerves, are occasionally affected simultaneously indicates that the disease is then not limited

<sup>1</sup> *Deutsch. Zeit. f. Nerven.*, 1900, xvii, 306.

<sup>2</sup> *Lancet*, 1893, i, 860.

<sup>3</sup> *Ibid.*, 1896, i, 1281.

<sup>4</sup> *Correspondenzblatt f. Schweizer-Aerzte*, 1902, xxxii, 366.

<sup>5</sup> *Revue de Méd.*, 1905, xx, 917.

to the sciatic but probably affects the lumbo-sacral plexus or the spinal roots. In the latter case there is usually an excess of lymphocytes in the cerebrospinal fluid, and according to Bonola<sup>1</sup> the fascia lata reflex, which is unaffected in sciatica, is lost.

**Diagnosis.**—The term sciatica is applied by custom to cases in which the symptoms are due to an organic affection of the nerve, as well as to those in which there is no evidence of structural disease. The first step must be, however, to separate these two classes, *simple sciatica* or *sciatic neuralgia*, in which there is spontaneous pain and tenderness of the nerve to pressure and to tension, but no pronounced sensory disturbance, degenerative atrophy of the muscles, or diminution of the Achilles tendon-jerk; and *organic sciatica* or *sciatic neuritis*, in which some or all of these signs of disease of the nerve are present. Some authors, as Gowers, regard all cases of sciatica with persistent tenderness of the nerve as neuritic, and consequently make simple sciatica or sciatic neuralgia very rare; but in facial neuralgia, in which there is certainly, as a rule, no disease in the nerve, its trunks may be quite as tender to pressure as the nerve in sciatica. Oppenheim and others have insisted that tenderness of the nerves is not a differential sign between neuritis and neuralgia. The pain of *hip disease* may radiate a short distance down the thigh; from sciatica it may be distinguished by the absence of tenderness in the nerve to pressure and tension, and the occurrence of pain on movement of the hip-joint, and on pressure on the trochanter. Disease of the *sacro-iliac synchondrosis* may be more difficult to recognize.

Although many cases of sciatica are due to a neuritic or morbid process in the nerve, the presence of marked sensory disturbance and degenerative atrophy of the muscles, with the absence of the Achilles tendon-jerk, should always raise the suspicion of more serious disease. If it is due to the compression of the nerve or its roots by a tumor in the pelvis, the sciatic trunk is not tender to pressure.

When the sacral roots are involved in *disease of the sacrum* or of the *cauda equina*, the symptoms are generally bilateral and more irregular in distribution, and the functions of the sphincters are, as a rule, affected; the pain is also generally referred to the cutaneous distribution of the affected fibres; the sensory or motor disturbances correspond to root distribution, and the nerve trunk is not tender.

In the early stages of *tabes dorsalis* the shooting pains may be limited to the sciatic distribution, but a careful examination of the case will generally reveal characteristic signs; in it the pain is almost invariably bilateral, and the nerves are not tender to pressure or stretching.

The pain of *intermittent claudication* occurs only after exercise, and is generally most intense in the distal segments of the limbs and is not limited to the course of the nerves.

**Prognosis.**—In simple sciatica this is good as regards ultimate recovery, but it is extremely difficult to predict the duration, although, as a general rule, it is proportional to the severity. Cases in which pain is

<sup>1</sup> *Rev. Neurol.*, 1912, xxiv, 324.

associated with signs of an organic nerve lesion are less favorable than the uncomplicated neuralgic cases. The outlook is less favorable in cases of long duration and where adequate treatment is not possible. Where the sciatic pain is due to some lesion extrinsic to the nerve which compresses or injures it, the prognosis is naturally dependent on the nature of the primary disease. Relapses are unhappily not infrequent.

**Treatment.**—The first essential in all cases is *rest*. Even in mild cases this should be made as absolute as possible for some days at least, and if it is adopted early severe cases may be often converted into slight ones. The patient should be confined to bed and all movements of the affected limbs should be restricted as far as possible, if necessary by a long splint. This treatment is applicable not merely to recent cases; obstinate and protracted cases which have resisted all other treatment often yield readily to it. It is often necessary to continue it four to six weeks, or even longer without break. Where it is not possible to obtain such complete rest an effort should be at least made to avoid all movements which give pain and cause stretching of or pressure on the nerve.

Constitutional conditions which are often predisposing causes should receive adequate treatment. In some of the acute cases the salicylates relieve the symptoms; iron and arsenic often prove useful in cases with anemia. The rectum should be emptied, and constipation avoided.

In acute stages the application of hot poultices along the course of the nerve may ease the symptoms. Counter-irritation by the application of the cauterizer or by blisters over the course of the nerve is very generally employed, and often seems to be of distinct value. Baths, douches, and especially the hot-air bath undoubtedly give relief and often influence the course very favorably, especially in its most chronic stages. When there is acute pain, symptomatic treatment may be forced into the first place. Antipyrine, phenacetin and such drugs often give relief for a time. Occasionally deep injections of morphine or cocaine into the nerve may be necessary, but they are only temporary measures and, owing to the nature of the disease, there is always a considerable danger of a drug habit. Morphine is the most effective, but cocaine in doses of from  $\frac{1}{8}$  to  $\frac{1}{4}$  of a grain may abolish all pain for hours.

Favorable results have been obtained by the injection of relatively large quantities of normal saline solution (50 to 100 cc.) into the sheath of the nerve. The injection is made in the upper part of the thigh; severe pain referred peripheralward, paresthesia, and muscular spasm of the limb, indicate when the needle enters the nerve. The acute pain disappears rapidly when the injection is commenced, which should be made slowly. More than two or three injections are rarely necessary, and often one is sufficient. Bum<sup>1</sup> recorded the result of this in 73 cases; he obtained a complete cure in 42, and 14 were much improved.

Half a gramme of antipyrine dissolved in an equal weight of distilled water has been also recommended as a local injection into the nerve. Excellent results have been also obtained by the injection of 10 to 15 cc. of physiological salt solution, or 1 per cent. cocaine or 4 per cent. stovaine, into the epidural space by Cathelin's method (Heile<sup>2</sup>).

<sup>1</sup> *Wien. med. Presse*, 1907, xlviii, 1660.

<sup>2</sup> *Neurol. Centralbl.*, 1912, xxxii, 596.

The galvanic current is often useful in the later stages; one large electrode should be placed over the nerve in the upper part of the thigh, the other on the leg over one of its branches or on the foot, and a constant current of 3 to 5 milliampères employed for five to twenty minutes.

It is rarely advisable to use massage in acute cases, but when the muscles become flabby or atrophy, it is of service, but pressure or tension of the nerve must be carefully avoided. Acupuncture may be tried; a series of six or more sterilized needles are thrust in to a depth of about two inches along the course of the nerve in the upper half of the thigh, and left there from twenty minutes to an hour. Many of the needles may pierce the nerve, but if they are inserted from above downward, only the first one causes much pain.

Nerve stretching has also fallen out of fashion, and probably rightly, but it may be necessary to have recourse to it in very obstinate cases. Bardenheuer has suggested cutting away the brim of the sciatic notch where the nerve passes over it, in order to leave the latter embedded in the soft tissues and free from pressure.

**Diseases of the Spinal Roots.**—The spinal roots may be affected by disease either inside the dura mater, or in their extrathecal course before they anastomose to form the plexuses; the lesions of the latter portions have been considered under the plexus lesions, and the diseases of the cauda equina are dealt with in another section. The disease of the roots may be primary, or the symptoms may be caused by compression or extension of disease from the surrounding parts.

Primary root lesions are extremely rare. One root alone may be injured by a tumor in the neighborhood of the vertebral column as frequently happens in malignant disease in this region, or spinal caries. Supernumerary ribs may produce an isolated palsy of the first thoracic nerve. But as Dejerine and his pupils<sup>1</sup> have pointed out, root lesions are more frequently secondary to intradural disease, and especially to compression and the invasion of them by a local tuberculous or syphilitic meningitis. A single root, either sensory or motor, only may be affected, but more commonly several roots are involved; there are frequently signs of associated disease of the spinal cord.

The distinguishing feature of all root lesions is the limitation of the symptoms to disturbance of the functions of the motor or sensory fibres of the root or roots affected; this distinguishes root lesions from lesions of the peripheral nerves, which almost invariably contain fibres of two or more roots. The onset, which may be either acute or slow, is generally with pain and paresthesia referred to the peripheral distribution of the sensory fibres involved; at this stage the skin of this region may be hyperæsthetic. The pain, which is generally at first paroxysmal but may become continuous, is, as a rule, very severe and of the same darting or shooting character as the pains of tabes dorsalis. If it is the intradural portions of the roots which are affected, sneezing or coughing may produce very acute pain in the affected root areas—*signe de l'éter-*

<sup>1</sup>Camus and Sézary, *La Presse méd.*, 1907, xv, 537; Camus, "*Les Radiculites*," Paris, 1908.

*nuement*—owing to the effect of the sudden increase of the intradural pressure on the irritable fibres. After a variable time the pain gradually diminishes, and the skin which was previously hyperesthetic become hypesthetic as the sensory root fibres degenerate or are destroyed. When only one root is involved this diminution of sensibility may escape notice owing to the considerable overlap of the adjacent root fibres; it is largely dissociative in character, the loss of pain sensation being, in contrast to the condition found after peripheral nerve lesions, more extensive than the insensibility to light touch (Head). Simultaneously with the appearance of hypesthesia, the muscles supplied by fibres from the affected ventral root or roots become paretic, and if the lesion is sufficiently intense they atrophy, and changes in their electrical reactions develop; but as almost all muscles receive fibres from two or more roots the paralysis of any muscle is rarely complete if the lesion is uniradicular. Cutaneous trophic changes, have been observed in the area of sensory distribution of the affected roots, probably in cases in which the root ganglia have been involved, and the ocular symptoms of irritation or paresis of the cervical sympathetic fibres may appear when the lower cervical and upper dorsal roots are affected.

**Diagnosis.**—This depends on the essentially radicular distribution of the symptoms. Spinal diseases, as tabes dorsalis, syringomyelia, and local lesions, may produce symptoms of this distribution, but these can rarely offer any difficulty. From local neuritis of a peripheral nerve it may be also distinguished by the fact that the nerve trunks are never very tender to pressure. When the primary disease is a meningitis which invades or constricts the roots, lumbar puncture may aid.

**Treatment.**—This should be directed to removing the cause; surgical intervention may be successful in the case of tumors, and if there are other symptoms of meningitis vigorous antisyphilitic treatment should be adopted if syphilitic infection cannot be excluded. In cases with persistent pain the intradural section of the dorsal roots may be necessary.

### NEUROMATA.

The peripheral nerves may be compressed and destroyed by tumors in the tissues through which they pass, or invaded by the infective granulomata, or by metastases of malignant tumors. The name *neuromata* was for long applied to all tumors seated in nerves, regardless of their histological structure; Virchow separated those which are composed of nerve substance proper, which he called *true neuromata*, from those which develop from the connective tissue of the nerves and contain no newly formed nerve fibres, *false neuromata*.

*True neuromata*, as defined by Virchow, are, however, exceedingly rare; they are found practically only in connection with the sympathetic system in the thoracic or abdominal cavities. They contain, as a rule, ganglion cells and non-myelinated nerve fibres. Although benign, they may infiltrate the surrounding tissues. They produce nervous symptoms only by compression of nerves, and are not tender to pressure.



Two cases have been recorded, by Knauss<sup>1</sup> and Kredel-Benecke,<sup>2</sup> in which there were multiple true neuromata in the subcutaneous tissues, but apparently most of the others described have been solitary. They are met with only in early life. Krauss<sup>3</sup> observed one in the lower lid.

*False neuromata*, which develop from the connective tissue of the nerves, are much more common. Their etiology is obscure; they generally occur in the first half of life, and in many instances they have been congenital. Occasionally they are hereditary; Petrén<sup>4</sup> found that in 10 of 60 cases of multiple neuromata there was definite evidence of the existence of similar tumors in the ascendant or collateral lines, while in 25 per cent. of all the cases there was an hereditary neuropathic taint. The appearance of neuromata has followed trauma of a nerve, but the false neuromata must be sharply separated from the bulbous swellings in the proximal ends of nerves which have been divided. Other cases have been attributed to infections and to arsenical poisoning.

**Pathology.**—As these tumors grow from the fibrous tissue of the nerves they are generally fibromata and of a benign nature, but they occasionally undergo myxomatous or cystic degeneration. More rarely they become sarcomatous; this is frequently observed when part of a tumor has been removed by operation or otherwise injured. Gliomata and lipomata have been described. The neuro-fibromata may originate from the epineurium, in which case the tumor generally lies on one side of the nerve and scarcely interferes with it, or from the perineurium or endoneurium; in the latter case it penetrates between and splits up the nerve into its separate bundles. The structure of the nerve fibres which pass through the tumor is, as a rule, unaltered, but there may be partial or complete disappearance of the myelin sheaths, while the axis-cylinders remain intact. Neuro-fibromata may be found on any of the spinal or cranial nerves, or on the intramedullary portions of the nerve roots. They vary in size from that of a pin-head to several inches in diameter; the smaller are generally spindle-shaped and elongated in the axis of the nerve from which they grow. There may be only one solitary growth on one nerve, or thousands may be distributed on all the somatic and visceral nerves; they are relatively common on the acoustic nerves and are then frequently bilateral.

**Solitary Neuromata.**—Symptoms are more frequently produced by solitary and isolated than by multiple neuromata, but many never cause inconvenience to the patient. The most prominent feature is pain which radiates peripheralward from the tumor along the nerve and its branches; it is generally intermittent, but may become continuous and so severe as to merit Smith's description of it as "agonizing torture." Movement of the limb and palpation of the tumor, which is generally tender to pressure, may induce an attack of pain, and, as in neuralgia, the influence of the weather may be sometimes observed. It has been known for long that compression of the nerve involved proximalward

<sup>1</sup> *Virchows Archiv*, 1898, cliii, 29.

<sup>2</sup> *Deutsch. Zeit. f. Chir.*, 1902, lxxvii, 239.

<sup>3</sup> *Zeitschr. f. Augenheilk.*, 1912, xxviii, 110.

<sup>4</sup> *Nordiskt Med. Arkiv*, Axel Key Festband, 1899.

to the tumor can relieve the pain. Cases have been described in which epileptic convulsions have been attributed to the irritative symptoms of neuromata, and in which the fits have ceased after their removal. Paresthesia occasionally occurs in the distribution of the sensory fibres which are affected, but loss of sensation and muscular paresis are rarely due to simple neuro-fibromata; reflex muscular spasms are sometimes seen. When palpable, the tumors are firm and evidently circumscribed, and they are movable in the transverse but not in the longitudinal axis of the nerve on which they are situated. Neuromata of the sheath of the optic nerve produce loss of vision, exophthalmos, limitation of the ocular movements, and pain in the eye; and tumors on any of the cranial nerves may give rise to the symptoms of complete or incomplete lesions of these nerves.

The only satisfactory *treatment* is removal of the tumors which produce inconvenient symptoms. This is rarely a serious operation, but resection of the nerve and primary suture of its divided ends may be necessary. Rapid growth, adhesion to or infiltration of the surrounding tissues, and evidence of acute destruction of nerve fibres indicate malignancy; amputation of the segment of the limb involved may be then necessary. The spontaneous regression of neuro-fibromata has been observed.

**Tubercula Dolorosa.**—Tubercula dolorosa, or the painful subcutaneous tubercles of Wood, may be either solitary or multiple; they are generally very small and lie directly under the skin. They grow slowly and are always benign. As a rule, they are neuro-fibromata, but are occasionally lipomata or sweat-gland adenomata which develop in connection with the smaller cutaneous nerves. They are generally exquisitely tender to pressure and may give rise to spontaneous pain. When troublesome they should be excised.

**Multiple neuromata or general neuro-fibromatosis** is a condition in which a large number of tumors is found on many or all the peripheral nerves, and even in the sympathetic system; R. W. Smith<sup>1</sup> recorded a case in which 450 were counted in one limb and over 2000 were present in the body. They are nearly always simple fibromata, but may become malignant. The condition is distinguished from the solitary neuromata, of which there may be more than one on a nerve, by the presence of a diffuse hyperplasia of the connective tissue of the nerves between the tumors, so that they can be felt or seen through the skin as thickened and beaded structures. They are rarely tender to pressure, and do not, as a rule, produce pronounced symptoms. Occasionally they give rise to local, or to vague and wandering pains, but they scarcely ever cause paralysis or serious sensory loss. Petré<sup>n</sup> observed one case, and collected others in which there was motor incoördination, probably secondary to loss of deep sensibility. These cases consequently resemble progressive interstitial hypertrophic neuritis. As a rule, inconvenience results only from the size of the tumors, although when seated on the spinal roots they may compress the spinal cord or the cauda equina, and within the skull they may give rise to the symptoms of intracranial

<sup>1</sup> *A Treatise on Neuroma*, Dublin, 1849; New Sydenham Society, London, 1898.

tumor. All forms of neuro-fibromatosis are frequently associated with cretinism, idiocy, or other mental deficiencies.

**Plexiform Neuromata.**—In this condition there is a diffuse hypertrophy of the connective tissue of all branches of a nerve, and the whole being embedded in fat or loose connective tissue gives rise to the appearance of a tumor. Occasionally they are pedunculated and hang in a sac of skin. They occur most frequently in the head and neck and are often congenital. They are not tender to pressure, and as they rarely produce nervous symptoms they belong rather to the domain of surgery.

**Molluscum fibrosum, or von Recklinghausen's disease,** is another form of neuro-fibromatosis which is characterized by numerous sessile or pedunculated cutaneous nodules which project from the surface. They are very variable in size, and may be either soft or hard, and are occasionally lobulated. The nodules are generally most numerous in the trunk and scalp, and are very rarely seen in the hands or feet. As von Recklinghausen first demonstrated, the nodules are neurofibromata of the terminal branches of the cutaneous nerves, but the fibrous tissue of the cutaneous vessels and glands may thicken and take part in their formation. There are rarely nervous symptoms, and the nodules are not tender. The disease is generally congenital, and is most frequently seen in degenerate types. It is often associated with nævi and skin pigmentation, and there are generally neuromata on the deeper nerves.

In the condition known as *elephantiasis neuromatosa* there is, in addition to a fibromatosis of the cutaneous nerves, a diffuse thickening of the skin, subcutaneous tissues, and even of the bone, of some part of the body, generally of one lower limb. Symptoms rarely result from affection of the nerves. The condition is usually congenital, and is frequently associated with pigmentation of the skin and other abnormalities. It may be distinguished from true elephantiasis by the absence of œdema and by the thickening of the nerves which are palpable.

### HERPES ZOSTER.

**Definition.**—Herpes zoster is an affection characterized by the appearance of erythema and cutaneous vesicles on the cutaneous distribution of the fibres of one or more dorsal roots.

**Etiology.**—The researches of Head and Campbell have made it probable that idiopathic herpes is an acute specific disease of the nervous system. This view is supported by the fact that it is occasionally epidemic and may have a seasonal prevalence, but no specific organism has been isolated. It occurs occasionally during the course of general infectious disease and in diseases in which there is an increased susceptibility to infection, as in general paralysis of the insane. It may also result from invasion of the dorsal root ganglia by tumors, or from the extension into them of a tuberculous process from spinal caries, or of inflammation from the meninges; it has been repeatedly observed in the course of acute cerebrospinal meningitis. In one case of herpes of the fourth thoracic segment the writer found a small metastatic abscess in the

corresponding intervertebral ganglion. Herpes has been attributed to general intoxications, including arsenical poisoning. It may occur in the early stage of acute poliomyelitis.

**Pathology.**—Barensprung, in 1861, first demonstrated changes in the intervertebral ganglia in a case of herpes zoster, but it was Head and Campbell<sup>1</sup> who definitely established the pathology. They showed that the herpetic eruption is always associated with acute interstitial inflammation of the dorsal root ganglia, or of their homologues on the cranial sensory nerves. In the earlier stages the affected ganglion is found infiltrated by small round cells; later part of the ganglion tissue is generally destroyed by the intensity of the inflammation, and many of its cells undergo acute necrosis; hemorrhages almost invariably occur in the inflamed area. The sheath also is generally involved, and the whole ganglion is swollen and hyperemic. When the inflammatory products are absorbed the disease may be represented only by a small area of scar tissue, or there may be cyst formation, but in the slighter affections there may be no permanent changes in the ganglion. The affection is not, as a rule, so localized as the eruption would indicate; in one case, although the eruption was limited to the third sacral root area, I found cellular infiltration not merely in the corresponding ganglion, but in all the neighboring ones of the same and of the opposite side. The inflammation may extend a short distance into the peripheral nerves or into the dorsal roots. When cells of the ganglion are destroyed their processes in the peripheral nerves and dorsal roots naturally undergo secondary degeneration. There is a striking similarity between the changes in the intervertebral ganglia in herpes and the affection of the spinal cord in acute poliomyelitis.

**Symptoms.**—Idiopathic herpes often sets in with fever and general malaise, which may last for three to five days, but the rise of temperature is, as a rule, slight. Occasionally, there are gastric disturbance and other constitutional symptoms, and enlargement of lymphatic glands has been observed. The eruption, which generally occurs on the third or fourth day of the disease, is often preceded by pain and hyperalgesia in the affected area. The skin becomes erythematous and finally vesicles develop, either while there is still fever, or as the temperature falls. The special characteristic of the disease is the limitation of the vesicles to the cutaneous distribution of one dorsal root, or more rarely of two or more adjacent roots. The eruption is not, however, uniform over this region, but is made up of a series of outbursts which follow the course of the small cutaneous nerves; there is a variable extent of overlap between adjacent herpetic areas. Herpes is much more frequent on the trunk along the course of the intercostal nerves than on the limb; according to Head those ganglia are most often affected which receive visceral sympathetic fibres. After a time the vesicles dry up and disappear, but their distribution is generally permanently marked by small, slightly depressed scars in the epidermis. On the face herpes is most common on the cutaneous distribution of the ophthalmic division

<sup>1</sup> *Brain*, 1900, xxiii, 353.

of the trigeminal nerve; when it occurs here vesicles may form on the cornea and lead to troublesome scarring and faceting.

The most prominent symptom is *pain* referred to the region of the eruption. The pre-herpetic pain which usually ushers in the attack is generally of a burning or stabbing nature; it may be extremely severe, but generally abates with the appearance of the rash. The post-herpetic pain or neuralgia is much more serious; it is also referred to the region of the eruption, but frequently spreads to adjacent segments; it is so severe and so intractable to treatment that it has driven the subject to suicide more than once. It occurs almost invariably only in old subjects. In a large proportion of the cases some loss of sensibility in the affected area follows an herpetic eruption, but in others sensation is unaffected. Petrén<sup>1</sup> has shown that there may be marked or even complete loss of pain and temperature sensibility, with tactile sensation unimpaired; and he has drawn attention to the fact that post-herpetic neuralgia occurs chiefly in those cases in which there is considerable or complete analgesia. The sensory loss is generally uniform over the whole of the affected area. A not uncommon complication is muscular paralysis of radicular distribution. Doucet<sup>2</sup> has collected 40 such cases. The palsy generally occurs in the neighborhood of the eruption, and is probably due to involvement of the ventral spinal root in the inflammatory process as it lies beside the affected ganglion; or occasionally perhaps to central lesions of toxic or infective origin. The facial nerve is perhaps the most frequently affected, generally in association with trigeminal or occipito-cervical herpes; it is, however, probably due to compression of the nerve in the aqueduct of Fallopius by a swollen geniculate ganglion which is simultaneously involved by herpes (Ramsay Hunt).<sup>3</sup> Paralysis of the ocular nerves has also been observed. In a few cases slight paraplegia has developed, as in a case published by Bruce;<sup>4</sup> it is probably due to extension of the disease to, or to the appearance of an independent focus in, the spinal cord.

**Treatment.**—The pain may be relieved by drugs in the earlier stage of the disease, but post-herpetic neuralgia is very intractable to all treatment; relief can be promised only by section of the dorsal spinal root corresponding to the ganglion affected, or by extirpation of the Gasserian ganglion where pain follows trigeminal herpes.

### MULTIPLE NEURITIS.

*Multiple neuritis, peripheral neuritis, and polyneuritis* are the terms applied to a group of diseases which are due to affections of the peripheral nerves, or rather of the peripheral motor and sensory neurones. These conditions are distinguished from paralysis limited to a nerve, or a group of nerves in close anatomical relation, by the fact that several nerves are affected simultaneously or in rapid succession (therefore

<sup>1</sup> *Zeit. f. klin. Med.*, 1907, lxxiii, 91.

<sup>2</sup> *Le Zona associé aux paralysies et aux amyotrophies*, Thèse de Paris, 1906.

<sup>3</sup> *Jour. Nerv. and Ment. Dis.*, 1907, xxxiv, 71.

<sup>4</sup> *Rev. of Neurology and Psychiatrie*, 1907, v, 885.

multiple neuritis), that the condition is always bilateral and more or less symmetrical, and that the longer fibres of the nerves which extend to the periphery of the limbs suffer more severely than the shorter fibres which are distributed to the proximal segments, or to the muscles and sensory structures of the trunk (therefore peripheral neuritis). The disease is characterized by the fact that it is limited to the peripheral neurones, while the central nervous system is intact, or any changes which may occur in it are merely a coincident effect and of no significance in the production of the clinical picture.

**Classification.**—A pathological classification is insufficient, as there are only two distinct types of pathological change; in the one the primary and essential process is a degeneration of the parenchyma or functional elements of the peripheral nerves; in the other it is an inflammatory or simple hypertrophic process of the connective tissue in which the fibres are ensheathed. But primary interstitial neuritis is extremely rare, and exists practically only as part of an infective disease—namely, leprosy—while the great majority of the cases are instances of parenchymatous neuritis, and it is impossible to subdivide this enormous group, as there are no essential differences in the neural changes in varieties due to different causes or characterized by special symptoms.

The classification by the clinical symptoms is equally unsatisfactory. These may consist only of motor phenomena, or of disturbance of sensation, or vasomotor symptoms may be the most prominent feature; but we rarely find that there are only motor, or sensory, or vasomotor symptoms; in fact, there is almost invariably disturbance of all the functions of the nerves affected, and affection of any of these functions may be the dominant symptom in different cases due to the same cause. It is, therefore, inadvisable to classify cases as of the motor, sensory, or vasomotor type.

An etiological classification is usually adopted and is the most useful, as it brings into prominence the cause of each case, and when multiple neuritis has been diagnosed the next step is to ascertain its cause. But an etiological classification cannot express the prominent features of a case, for though certain poisons may have an affinity for nerve fibres with certain functions, such a selective action is not always apparent.

**Etiology.**—The wide distribution of the affection and its symmetrical regularity indicate, as Gowers points out, that multiple neuritis must be directly due to an altered blood or lymph state which has equal access to all parts of the body, and, indeed, its cause is invariably some poison which is introduced into the body from without, or produced within it by infective microorganisms or by a derangement of normal metabolism. Many of these poisons, especially those of inorganic nature and of simple chemical constitution, seem to have a selective action on the peripheral nerves and may produce disturbance of function in no other organ of the body; motor palsy, for instance, may be the only symptom of chronic lead poisoning. But in the majority of the cases which are due to bacterial toxins or to poisons of complex chemical nature the symptoms of multiple neuritis are only part of the result of a general infection or

intoxication, or, as is the rule in the acute specific fevers, the neuritis may appear when the symptoms of the infection are on the wane.

There is a group of infective diseases in which a peripheral neuritis may be the only or the most prominent symptom. Beriberi is the best known example, but there are undoubtedly other, as yet ill-defined or scarcely recognized, infectious conditions which may manifest themselves only by the symptoms of a primary multiple neuritis.

It not infrequently happens that there are two or more factors at work, predisposing as well as directly exciting causes. Neuritis occasionally develops in the course of pulmonary tuberculosis, and although it is probable that it may be at times due to the action of the tuberculous toxin alone, it is very rarely that the influence of alcohol can be excluded. Similarly, quite a series of cases of neuritis have been observed after the administration of phosphate of creosote to patients with phthisis;<sup>1</sup> here the tuberculosis was probably the predisposing, the drug the exciting, cause. In the extensive epidemic of peripheral neuritis in the north of England, which was caused by the contamination of beer with arsenic, it is probable that there were also two distinct but coincident etiological factors, namely, alcohol and arsenic. Finally, there can be no doubt that most marasmic and cachectic conditions make the nerves unduly vulnerable to noxious agents.

1. **Neuritis Due to Poisons Introduced into the Body from Without.**—These poisons may be either: (a) *Metallic or inorganic substances*, as lead, arsenic, mercury, copper, carbon monoxide, or bisulphide of carbon; or (b) *organic substances*, as alcohol, dinitrobenzene, various aniline compounds and derivatives, sulphonal, and other drugs; finally, the poison may be of more complex constitution, as the toxins of ptomaine poisoning. Neuritis due to any of these causes, except perhaps carbon monoxide, is generally associated with chronic intoxication and rarely follows a single administration of the poison, no matter how large the dose.

2. **Neuritis Secondary to, or Associated with, Diseases Due to Toxins Produced Within the Body.**—These toxins may result from: (a) *Bacterial infections*, most frequently diphtheria, smallpox, typhoid, and scarlet fever. The toxins produced by septic processes may also give rise to a multiple neuritis, as is the case in puerperal neuritis; (b) *derangement of metabolism* as in diabetes and other constitutional diseases. There is very little evidence that gout produces a polyneuritis, but rheumatism may certainly be a cause (Schulhof).<sup>2</sup> As a rule, these conditions act more as predisposing factors than exciting causes.

3. **Toxins Produced within the Body Which Have a Primary Action on the Peripheral Nerves.**—A variety of neuritis which sets in with malaise and fever and more or less severe constitutional symptoms is not infrequently observed, and probably comes into this group (acute febrile polyneuritis). It is often called post-influenzal, and although the influenza bacillus or its toxins may be occasionally the exciting cause, it seems doubtful if it is often so. These cases are distinguished by the severity of the neuritis as contrasted with the slight constitutional

<sup>1</sup> Huet, *Neurol. Centralbl.*, 1907, xxvi, p. 60.

<sup>2</sup> *Med. Klinik*, 1913, ix, 952.

disturbances which precede its onset, or the complete absence of prodromal symptoms. Occasionally exposure to cold is the only apparent exciting cause in cases which probably belong here.

4. **Neuritis associated with cachexia or malnutrition**, such as occurs in anemia, tuberculosis, syphilis, malignant disease, and in old age. These may be directly due to intoxication, or in some cases, as in tuberculosis, to the action of the bacterial toxins which produce the cachexia. In other cases the general malnutrition merely leads to an increased vulnerability of the nerves; or the nutrition of the nerves may be directly interfered with, as in severe peripheral arteriosclerosis.

5. **Neuritis Due to the Invasion of the Peripheral Nerves by Bacteria.**—Leprosy is the classical example of this type. Nerves involved in septic wounds may be invaded by pyogenic bacteria which occasionally extend along the trunk and may involve adjacent nerves—*ascending neuritis*—but the picture of multiple neuritis can scarcely arise. Gonorrhœa may also lead to interstitial multiple neuritis (Frisco), or to acute neuritis either with or without arthritic disease, and Eberth's bacillus has been found in the nerves in typhoid neuritis.

In certain cases it is impossible to determine the direct cause, as in the neuritis of pregnancy (Dustin<sup>1</sup>) and that which occurs associated with gastric ulcers (Klippel and Pierré—Weil<sup>2</sup>).

**Incidence.**—Multiple neuritis is essentially a disease of adult life. It occurs rarely in childhood, and then practically only after the acute specific fevers, especially diphtheria. It may follow the treatment of chorea with "heroic" doses of arsenic. Polyneuritis is also uncommon in advanced age, and when it occurs is usually slight in degree. Hereditary influences are rarely observed. The disease is more common in females, probably due to their greater liability to alcoholic neuritis.

There are many factors which may determine the localization or incidence of the disease. Perhaps the most important of these is the effect of overwork or exhaustion, especially when the nutrition is subnormal or when the metabolic equilibrium is otherwise upset. Thus Edinger explains the predominant paralysis of the extensors of the wrist and fingers in lead palsy in painters; in composers, on the other hand, who not infrequently suffer with lead palsy, the paralysis is often limited to the small hand muscles with which they chiefly work. Occasionally the mode of the infection determines the type of the palsy; the soft palate is generally the first organ affected in diphtheritic neuritis following pharyngeal diphtheria, while the abdominal muscles were the earliest and most severely paralyzed in a few cases in which the seat of infection was the umbilicus in infants.

**Pathology.**—The changes in multiple neuritis differ from those of local neuritis not only in their distribution, but also in their nature. In the latter, the primary changes affect the interstitial tissues, and are generally of an inflammatory nature; the condition produced is consequently a local interstitial neuritis. Multiple neuritis is the result of degenerative, parenchymatous changes in the peripheral nerves.

<sup>1</sup> *Now. Iconographie de la Salpêtrière*, 1909, xx, 349.

<sup>2</sup> *L'Encephale*, 1909, iv, 417.



The amount of structural change is very variable; it depends not only on the intensity of the disease, but also on its cause and nature, and particularly on its duration. To the naked eye there is rarely any definite abnormality, although in the acuter cases in which the primary disease has led to a reaction in the interstitial tissues, the nerves may appear red and swollen, and there may be a distinct hyperemia of their sheaths. These changes, when present, are generally most distinct in the smaller peripheral branches. In chronic cases, the nerves may be reduced in size and unusually firm and opaque. Microscopic examination shows that the myelin sheaths are broken up and disintegrated into irregular masses which stain black with osmic acid; in cases of longer duration this fatty degeneration product may have been already removed. In the early stage the axis-cylinders may be intact; more commonly they are swollen and varicose and stain feebly, or if the neuritic process is further advanced they may be broken or completely disintegrated. The protoplasm around the nuclei increases in amount and penetrates between the fragments of the disintegrating myelin, and the nuclei proliferate by direct division. Some of these neurilemmal nuclei form independent cells which assume phagocytic functions or become fibroblasts and lead to the formation of an excess of connective tissue; but others fuse end to end to form those nucleated strands of undifferentiated protoplasm into which, when regeneration commences, the new outgrowing axis-cylinders penetrate.

Even in parenchymatous neuritis there is frequently some evidence of inflammatory or reactionary changes in the interstitial tissue; the vessels become congested and there is often some serous effusion, and occasionally a small round-cell infiltration.

These neuritic changes differ from secondary or Wallerian degeneration of nerve fibres in many important particulars. In the first place, they are always most advanced in the most distal segments of the nerves—multiple neuritis has been consequently aptly called peripheral neuritis—and the intensity of the changes always diminishes steadily proximalward; indeed, it is the rule that except in the severest cases the larger nerve trunks, as the sciatic, contain a much smaller proportion of degenerated fibres than their peripheral muscular or cutaneous branches, while in the dorsal and ventral spinal roots there may be no degenerated fibres. Secondly, the morbid changes are much less uniform than in fibres which are undergoing secondary degeneration. In almost all cases of toxic parenchymatous neuritis there are patches of that form of segmental periaxial neuritis which was originally described by Gombault, in which the disease is limited, or practically limited, to a segment of the myelin sheath; in its slighter forms the axis cylinder is not interrupted and the portion of the fibre distal to the lesion does not undergo secondary degeneration, and may be perfectly normal. Numerous transition stages may be found between these local lesions and the diffuse degenerative changes. Thirdly, not only is the affection of any one fibre irregular and diffuse, but the number of fibres affected in any nerve is extremely variable; even in the severest cases it is rare to find all the fibres of even the smaller nerves degenerated.

The interstitial changes are not without importance; the extreme tenderness of the nerves has been ascribed to this, but it seems extremely doubtful if it is so, as the affected nerves are almost invariably tender to pressure, while the presence of such inflammatory changes is very inconstant. When, however, they are pronounced they may lead to the formation of an excess of connective tissue, which ultimately replaces the degenerate fibres and may offer a serious obstacle to their regeneration, or may even interfere with the vitality of those which were originally unaffected.

Changes are occasionally present in the central nervous system; when the dorsal roots are involved degenerated fibres will be naturally found in the dorsal columns of the spinal cord; but multiple neuritis is essentially due to toxic causes, and the dorsal column fibres are extremely liable to degenerate in all forms of general intoxication. More important and much more constant are the changes in the large ventral horn cells; they were present in some degree in every case of multiple neuritis examined by the writer. They may be described as chromatolysis of the cells. Similar changes may be generally found in the cells of the dorsal root ganglia, and not infrequently in other parts of the central nervous system, as in the cerebral cortex; the association of mental disturbances with multiple neuritis may be the clinical manifestation of the affection of the latter region (Bonhoeffer). Small inflammatory foci and minute hemorrhages have been described in the spinal cord, but have been much too slight and inconstant to play any part in the production of the symptoms. They can be only attributed to the action of the toxin.

The nature of the marked alterations in the large motor ventral horn cells, and their relation to the neuritic changes, has been much discussed. There are evidently three possibilities: In the first place that the cell changes are the primary effect of the action of the noxious agent which produces the neuritis, and that the degenerative lesions in the nerve fibres are secondary to the affection of their trophic centre; this hypothesis seems scarcely tenable, as there is no constant relation between the cellular and fibre lesions; in fact the former are occasionally absent, and it is well known that the cell disease must be very acute to produce an acute degeneration of its axis cylinder. Further, the nerves are so diffusely and irregularly affected, and the disease is so frequently limited to their distal extremities that it is extremely improbable that it can be secondary to an acute cell lesion. In favor of this view it has been argued that the portions of the neurone most distant from their trophic cells must be the first portions to degenerate when the vitality of the cell is lowered. Even though this view may not give sufficient support to the hypothesis that the cell changes are the primary lesions in multiple neuritis, it explains very satisfactorily the chief incidence of the disease on the peripheral segments of the nerves.

The second possibility is that the cell changes are secondary to the degeneration of the fibres—that they are comparable to the retrograde chromatolysis which may be seen in any cell after its axis-cylinder has been cut across. This may be an important factor, but it is improbable that it is the only one, as it must be recognized that there is no close

parallel between the neural and the cellular changes, and that the cellular alterations do not always conform to the type of retrograde chromatolysis. The third possibility, and the most acceptable, is that all portions of the peripheral motor and sensory neurones are simultaneously affected, but the most prominent alterations are found in their least resistant portions, namely, the segments of the fibres furthest removed from the cell body.

The importance of the muscular changes must be remembered. The degree of change depends on the severity and the acuteness of the neuritis, but in addition to the degenerative or regressive changes, such as succeed section of a nerve, there is also often fatty, granular, or waxy degeneration of the muscle fibres, a result of the direct action of the toxin. There is frequently, too, evidence of inflammatory processes in the interstitial tissues of the muscles, an interstitial myositis.

**Symptoms.**—It seems advisable to describe fully one form in which all symptoms may be found in various combinations; other varieties of the disease will then need only short references.

*Alcoholic neuritis* is much the most frequent form—in Great Britain, at least, it is probably much more common than all the other varieties put together, and may be taken as a type. In most cases all the functions of the peripheral nerves are affected, but occasionally the symptoms are limited to either motor or sensory disturbances, or incoördination of movement may be the most prominent feature; the latter type has been called *neuro-tabes peripherica*, or alcoholic pseudo-tabes. Alcoholic neuritis occurs practically only in middle adult life, but a few cases have been recorded in the first decade; in these the disease has generally developed after a single large dose of the poison. In England and America it occurs much more frequently in women than in men, according to Gowers in about the proportion of 3 to 1; in Germany, from the statistics of Remak, it seems to be more common in males, The social habits of the people must largely determine its incidence, but women undoubtedly seem more vulnerable to the poison.

It usually results from the use of the stronger forms of alcohol, especially from spirits, but it is seen not infrequently in beer drinkers, and especially in constant and steady tipplers. Undoubtedly, it affects chiefly those who habitually take a considerable amount of alcohol, but are rarely drunk; occasionally, however, an attack may follow a severe bout of drinking or delirium tremens. There is frequently some exciting or predisposing cause in addition to alcohol, as a general infection or insufficient nutrition; pulmonary tuberculosis is found in a considerable proportion of the cases, at least of those which come to a fatal termination. Lindl<sup>1</sup> found some evidence of local or general neuritis in 214 of 300 drunkards. The slighter forms often escape recognition.

**Onset.**—In some cases premonitory symptoms occur which may be either the result of the alcoholic intoxication or a part of the disease. To the former group belong general constitution disturbances, as morning vomiting and other symptoms of disorder of the digestive system, or

<sup>1</sup> *Der Alkoholismus*, 1093, Neue Folge, Heft i.

dyspnœa and symptoms of affection of the circulatory system. In other cases loss of memory or slight mental impairment may be the first indication that anything serious is wrong. But the true prodromata, when there are any, are numbness and tingling of the fingers and toes, vasomotor disturbances in the extremities, painful cramps of the muscles, especially in the calves, and vague pains. These are undoubtedly part of the disease and constitute its first symptoms, but as they often exist for long periods before the onset of classical signs, it is important to recognize them as premonitory of a more serious affection. In this early stage there is often some fever.

The onset of the *neuritis* is nearly always gradual, often slow and insidious; as the patient's memory is frequently impaired, the earliest symptoms may be overlooked or attributed to the effects of drinking, and it may be difficult to ascertain the duration or the mode of onset. An acute onset is rare, but cases have been recorded in which a sudden loss of power in the limbs apparently occurred. But these acute attacks are generally more apparent than real; they are usually seen only after acute illnesses or a bout of drinking, during which the earlier symptoms were not observed. In the common forms the first definite symptom is usually disturbance of sensation with paresthesia in the hands and feet, vague diffuse pains which are often very severe and always increased by movement, and tenderness of the muscles to pressure; these are soon followed by motor symptoms. The symptoms are almost invariably bilateral and symmetrical from the onset and more prominent in the lower than the upper limbs, but in acute cases one limb or the functions of one nerve may be unduly affected at first. The further evolution is variable; in the severer forms the patient is generally bedridden in a few days, but, as a rule, the disease progresses more slowly and reaches its acme in three or four weeks. Occasionally its course is much more chronic; it may progress slowly for months in mild cases when alcohol is not withheld. The ataxic variety has often this slow and chronic course.

**Motor Symptoms.**—Paralysis may involve either the upper or lower limbs, more commonly both, but it is invariably more marked in the distal than in the proximal segments. The legs are almost always more affected than the arms, often they alone are involved. The trunk muscles escape frequently, and any considerable degree of palsy in the muscles supplied by the cranial nerves is rare. It must always be borne in mind that true muscular paralysis may be simulated by the reluctance of the patient to perform movements with the full amount of power which is possible to him, owing to the pain which forcible contraction of the tender muscles produce; a careful examination of the electrical reactions of all muscles may be necessary to ascertain the extent of the affection. In the usual type the first symptom is a slight difficulty in walking, especially on uneven surfaces or in ascending stairs, owing to the patient's inability to raise the toes from the ground. Dorsiflexion of the feet is very feeble or impossible, and the foot when unsupported hangs in the extensor position with its dorsum almost in the line of the axis of the leg. With this, some malposition of the toes is generally associated; in the earlier stages they are generally hyperextended at their distal and flexed

at the interphalangeal joints, except the great toes, which are often extended at all their joints; later all the toes are generally fully flexed and curled into the soles.

To avoid the constant danger of the toes catching in any obstacle, or even tripping him on level ground, owing to the ankle-drop, the patient raises his feet unusually high by flexion of the hips, and throws them forward with each advancing step; the gait has been consequently called *steppage gait*. It is almost characteristic of peripheral neuritis. It occasionally happens that all the muscles of the anterior tibial groups are not equally affected—the tibialis anticus often escapes when the peronei and extensors of the toes are paralyzed.

The symptoms of palsy are, however, very rarely limited to the muscles on the front and outer aspects of the legs; those which are usually next affected in both time and degree are the calf muscles. Walking is then more seriously interfered with, as the patient cannot spring from his toes or even fix the ankle-joints properly. The muscles of the thighs and hips suffer later and, according to the general rule, being proximal muscles, to a considerably less degree; indeed, the muscles which move the hip-joints are much affected only in the severest forms, and by that time the patient is invariably bedridden. The paralyzed muscles are soft and flabby to touch, and evidently hypotonic; when the paralysis has lasted more than two or three weeks they begin to waste, and the atrophy corresponds approximately to the degree of the paralysis. In the latter stages, however, when the atrophied fibres become replaced by connective tissue, the muscles may be firm and fibrous to touch.

In the upper limbs the paralysis, as a rule, first expresses itself by affection of the extensors of the wrist and fingers. Rarely, however, does the extent of the paralysis coincide with the nerve distribution; in the arm the supinator longus and the extensor ossis metacarpi pollicis often escape, or are less affected than the long extensors. This type of paralysis and the wrist-drop that results, although most characteristic of lead palsy, is not infrequently seen in alcoholic neuritis. The loss of power in the extensors of the wrist and fingers leaves the arms almost useless; the patient cannot grasp any object firmly owing to the mechanical disadvantage at which the flexors of the fingers must work when the extensors of the wrist are paralyzed. But the flexors of the wrist and fingers rarely escape, and in the severer cases may be as powerless as the extensors. Occasionally the intrinsic muscles of the hand are affected early; according to Gowers those of the thenar and hypothenar groups always suffer later than the interossei. Of the muscles above the elbow, the triceps generally suffers more severely than the biceps, while those which move the shoulder-joints are rarely much affected. In the atrophic stage the wasting is generally most prominent on the dorsal surfaces and ulnar borders of the forearms; in the hands it may be indicated by flattening of the thenar and hypothenar eminences and by hollowing of the interosseal spaces.

The muscles of the trunk are involved in severe cases, most commonly those of the abdominal walls; this impairs the power of expelling the contents of the bladder and rectum, and coughing and sneezing. If the

flexors of the thighs are weak as well as the recti abdominis, the patient becomes unable to raise himself from the recumbent position. There is considerable palsy of the muscles of the back and neck only in the severest cases. Paralysis of the diaphragm is occasionally observed; respiration is consequently severely implicated, as the intercostal muscles rarely escape completely, and a fatal termination is therefore probable.

The functions of the cranial nerves are sometimes involved; lingual, palatal, or pharyngeal palsies may develop in very severe cases, generally when they are progressing to a fatal termination; laryngeal paralysis is very rare. The tachycardia, which is often a prominent symptom, is generally attributed to disease of the pneumogastric nerves; Dejerine has demonstrated parenchymatous degeneration of them.

Bilateral facial palsy, which is not infrequently seen in infective multiple neuritis, is rarer in the alcoholic form. So also are ocular palsies, though many cases in which they have occurred have been recorded; they are probably due to hemorrhagic polioencephalitis superior (Wernicke's form), which develops most often in the subjects of chronic alcoholism, and not to a neuritic affection of the ocular nerves. Nystagmus, however, is not infrequently observed. The pupillary reactions are very rarely altered, but the Argyll-Robertson phenomenon has been observed in alcoholic neuritis and in chronic alcoholism (Nonne<sup>1</sup>). Optic neuritis has also been described.

The state of the *electrical reactions* of the nerves and muscles is very variable. The excitability of the nerves which are affected diminishes rapidly and generally disappears completely; it is interesting that little or no reaction may be obtained from nerves the functions of which are almost intact; this is evidently the result of incomplete lesions. No contraction can be obtained, as a rule, by the direct application of the faradic current to muscles which are much paralyzed; in the less affected the reaction may be incomplete, as a small proportion of the fibres may respond. With the galvanic current all forms of the reaction of degeneration may be obtained, but the current necessary to produce any contraction is generally greater than normal. In severe cases the muscles often eventually lose their galvanic excitability.

Although paralysis is invariably the most prominent motor symptom, there is occasionally evidence of motor irritation or hyperactivity. The tonic spasms or cramps of calves and other muscles which occur as frequent premonitory symptoms occasionally persist in slighter cases throughout. Fibrillation, too, is occasionally seen, and irregular tremor of the hands, tongue, and facial muscles is a well-recognized symptom. These phenomena are probably due to irritation of undegenerated fibres. Still more important is the reflex or voluntary contraction of relatively intact groups of muscles, by which the limbs are held in the positions which are most comfortable, and by which the patient resists passive movements which give him pain. Thus the knees are almost always semiflexed by contraction of the hamstring muscles; the shortened muscles may become permanently contracted.

<sup>1</sup> *Neurol. Centralbl.*, 1912, xxxi, 6..

**Sensory Symptoms.**—The tingling, numbness, and other subjective sensations in the distal segments of the limbs generally persist throughout the course but the pains which are an almost constant symptom are a more serious cause of suffering. They may be aching, burning, or shooting in character, and are often very severe. They are generally more diffuse and less intermittent and lancinating than those of tabes dorsalis, and, as a rule, are referred to the deeper tissues of the limbs or joints, rarely to the course of the nerves or to the skin. They may occur spontaneously, but are excited or increased by movement, either active or passive, of the limbs; severe paroxysms often occur during the night and deprive the patient of sleep. Not infrequently there is constant and very troublesome neuralgic pain in the soles of the feet.

But the most prominent and constant sensory symptom of multiple neuritis is the extreme *tenderness* of the muscles to pressure; this is probably never absent and is almost a pathognomonic sign. It is usually greatest in the calves and in the regions in which there is considerable paralysis, but muscles which are in no degree paralyzed may be very tender. In the severer cases the limbs cannot be touched without eliciting cries of pain, and even the weight of the bed-clothes may be intolerable. It becomes agonizing for the patient to lie long in any position, and yet he dreads being moved, owing to the pain which any change of posture produces. The affected nerve trunks are also often very sensitive to pressure, but this is neither so constant nor so prominent as the muscular tenderness. Occasionally the superficial nerves may be felt slightly enlarged or swollen.

Objective disturbances of sensation are nearly always much less marked than the motor palsies, but they are rarely quite absent. They are most marked in the distal segments of the limbs and diminish gradually proximalward, ceasing in the legs, as a rule, about the middle of the thighs and rarely marked in the arms above the elbows; but there is never a definite boundary line between the normal and anesthetic skin, and it is characteristic of the hypesthesia that it does not correspond in extent to the distribution of either the peripheral nerves or the dorsal roots. Usually the most definite loss is to light touches, which may not be perceived in the feet or legs and felt only unnaturally on the thighs. Sensibility to pain is less commonly lost, but there is not infrequently some delay in perception of painful stimuli. The appreciation of temperature also may be diminished or absent in the extremities, but this is far from constant even in the severer cases. But more characteristic of neuritis is a curious combination of hyperesthesia and hyperalgesia; in areas of skin which are more or less insensitive to light touches a pin-prick or a scrape may cause intense pain, and even washing or rubbing the skin may give rise to much discomfort. Occasionally the sense of position is more affected than cutaneous sensibility.

Another symptom which results from loss of sensation, but expresses itself by disturbance of motion, is *ataxia* or incoördination. It is not frequent although occasionally it may be the most prominent feature. Incoördination becomes a prominent symptom when the afferent nerves from the muscles and joint are unduly affected. Dejerine has given

this condition the name *neuro-tabes peripherica*, owing to the similarity of its symptoms to those of *tabes dorsalis*; but in his two original cases, as well as in others, the dorsal spinal roots and the dorsal columns of the cord were intact, and the disease was limited to the peripheral nerves. The ataxia is generally apparent only in the movements of the lower limbs, and especially in walking. The gait may closely resemble that of *tabes dorsalis*, and the similarity is increased by the fact that the patient is more unsteady in the dark or when his eyes are closed. Ataxia of this type must be distinguished from the disorders of movement which may result from paresis alone; it may be observed in cases in which the muscular power is intact or relatively little affected. In some cases the ataxia precedes the development of paralysis; in others it may remain the chief symptom. In the latter class it often sets in rapidly, and is usually accompanied by at least some diminution of cutaneous sensibility and frequently by severe pains and muscle tenderness. The sense of position and of movement is diminished in the majority of these cases.

**Reflexes.**—Loss of the tendon reflexes—including the knee- and ankle-jerks in the lower limbs, the biceps and triceps jerks in the arms—is one of the cardinal signs, and the most delicate test we possess of disease of the peripheral nerves. The knee-jerks may be absent not only when there is definite palsy or loss of sensibility, but also in chronic alcoholism when there is yet no suspicion of neuritis. Their absence is, however, not an absolute rule; they persist in rare cases, despite the presence of considerable paralysis, and they have been found definitely exaggerated in the early stages. This is difficult to explain; one theory assumes that it is due to the co-existence of a spinal lesion, probably also the result of the intoxication that produces the neuritis; another attributes it to a state of excessive irritability of the sensory fibres which leads to such an exaggeration of the afferent impulses to make it possible for them to overcome the obstacle offered by a slight degeneration of the motor fibres of the reflex arc. Absence of the tendon-jerks is often the most persistent sign of neuritis in cases which improve; they may not reappear for years after complete disappearance of the other symptoms. The cutaneous reflexes are generally diminished or abolished, but they occasionally persist, even when there is considerable motor palsy, especially in cases with marked hyperalgesia. The plantar reflexes, when they do not disappear, are invariably of the flexor type.

**Sphincters.**—The functions of the bladder and rectum are, as a rule, unaffected, although in cases in which there is much mental disturbance the patient may pass urine involuntarily or have symptoms of retention. Occasionally, however, dysuria and retention have been observed; such sphincter affection has been generally attributed to the co-existence of spinal disease, but this has not been demonstrated, and it is possible that the neuritic affection may extend to the nerves of the bladder.

**Trophic and Vasomotor Changes.**—Apart from the atrophy of the paralyzed muscles trophic changes are not a prominent feature. In chronic cases the skin, especially of the hands and fingers, loses its natural wrinkles and becomes thin and shining, but true “glossy skin,” which is frequently seen after severe local nerve lesions, is rarely met with in multiple neu-



ritis. That of the legs is generally dry, rough, and covered with scales of desquamating epidermis. The hair and nails become dry and brittle, and the latter may be furrowed longitudinally and may grow irregularly. Bedsores are rare, and severe trophic changes are almost never seen.

Of the vasomotor change, *œdema* is the most prominent; it occurs in a considerable proportion of the severer cases. It is generally localized and is most common on the dorsa of the hands and feet, and about the ankles. In the majority of the cases it certainly develops independently of vascular and renal disease, and is probably due to an affection of the vasomotor fibres. Sometimes it does not become apparent until the patient begins to walk or to sit up with his feet dependent; in other cases it may be an early or the initial sign, as it so frequently is in beriberi. Local anemia is occasionally observed, and Ross has reported a case of alcoholic neuritis with the symptoms of Raynaud's disease. Profuse sweating is common in the early and acute stages, especially in the extremities of the limbs.

Several cases have been recorded in which neuritis set in with or immediately after an attack of polyarthritis; most probably the two conditions were here due to the same cause, but in other cases one or more joints have been observed to swell and become painful during the course of neuritis; the changes may ultimately produce ankylosis. More chronic arthritic changes, with pain and peri-articular thickening and ultimately limitation of movement, are not uncommon.

**Mental Symptoms.**—Psychical disorders are so common in multiple neuritis, especially in the alcoholic form, and so frequently of the same type, that they may be regarded as part of the disease; in fact some authorities speak of these mental symptoms as the polyneuritic psychosis. Korsakow first carefully analyzed and described them in their most common form (see Vol. II, p. 427).

**Course.**—This is very variable; it depends largely on the cause, more on the stage in which the patient comes under treatment and when the action of the poison is arrested. As a rule, the more severe and acute the symptoms the longer will be their duration; they generally increase for a time after the alcohol has been stopped, and then remain stationary until improvement sets in. But this can be rarely expected within one or two months. The first sign of improvement is usually diminution of the pains and cutaneous hyperalgesia, but tenderness of the muscles often persists until recovery is almost complete. The disturbances of cutaneous sensibility generally disappear before there is much return of power. This occurs first in the muscles which were last and least affected, that is, in those which move the proximal joints of the limbs, and progresses distalward, but only very slowly; in even the most favorable cases it requires months for muscles that were definitely paralyzed to regain their functions. On the other hand, complete return of power in all muscles may be expected eventually in almost all cases which improve, provided that the patient does not revert to the abuse of alcohol. As the paresis disappears the wasted muscles slowly regain their bulk, but may not attain their normal size for months. The most serious obstacle to complete recovery is the shortening and contracture of the antagonists

of the most affected muscles. It is very frequently seen in the calf muscles; the ankle is then more or less fixed in the extensor position and the foot cannot be dorsiflexed, so that the patient is unable to get his heel to the ground on attempting to walk. The absence of the tendon-jerks, and alterations in the electrical reactions are usually the last persisting evidence of the disease in these favorable cases.

The course of alcoholic neuritis is not always so favorable. In cases with acuter onset, in which the limbs become powerless within one or two weeks, the paralysis may spread to the muscles of respiration and produce a fatal result; and not infrequently death occurs from cardiac failure in the acute stage of the disease, owing either to paralysis of the vagi nerves, or to direct affection of the heart muscles. Cases have been recorded which have come to a fatal termination within a week, but probably most of these were cases of Landry's paralysis.

The chief complications are the effects of alcohol on other organs, as the heart, liver, and gastro-intestinal tract. Pneumonia and bronchitis, which occur not infrequently, are serious complications. Pulmonary tuberculosis is perhaps the most frequent of the serious complications and probably the most common cause of death. Its relation to the neuritis may be twofold; on the one hand, the tuberculous toxin may be an etiological factor, especially when combined with other causes, as with alcohol; on the other hand, alcoholic subjects are very liable to tuberculosis, or latent foci may become active when the patient is confined to bed with neuritis.

**Diagnosis.**—When the essential features are recognized this can present no difficulty in the majority of cases. The wide distribution of the affection of both motor and sensory functions, its rigid symmetry, and above all its greatest intensity in the distal segments of the limbs, characterize the disease in the typical cases. It is, however, very important to remember how variable the symptoms may be in different forms, and even in cases due to the same cause.

The condition with which it may be most easily confused is perhaps *acute poliomyelitis*; it is little over a quarter of a century since a definite distinction was drawn between them. The difficulty is chiefly present in cases which occur in early life, as then multiple neuritis is relatively rare, while infantile palsy is common. The onset of poliomyelitis is, however, more acute, and is generally accompanied by severer constitutional disturbances; the paralysis is rarely symmetrical, and is often more marked in the proximal than in the distal segments of the limbs; further, complete paralysis of the abdominal and trunk muscles is not uncommon, while in multiple neuritis it is exceptional. Sensory disturbances and the severe and persistent neuritic pains are absent, and although the paralyzed limbs may be at first painful to pressure or movement this soon disappears and the nerve trunks are not unduly tender. Neuritis is generally progressive for a few weeks, while poliomyelitis usually comes to a standstill in a few days and the muscles least affected recover quickly; then the irregular and random distribution of the paralysis is evident.

The differential diagnosis between multiple neuritis and *Landry's*

*paralysis* is more difficult. The course of the latter is more acute, and, as a rule, death results within ten days or a fortnight; the constitutional symptoms are generally slight and the sensory phenomena are limited to pain in the back and occasionally slight numbness in the extremities; definite anesthesia does not occur, and neither the nerves nor muscles are tender to pressure. But its most characteristic feature as contrasted with polyneuritis is that the muscles of the trunk and of the limbs are equally affected, and that the paralysis ascends the trunk from the legs to the arms, and does not appear in the hands and feet simultaneously or successively and involves the musculature of the trunk last, as does multiple neuritis. Finally, the paretic muscles rarely waste and the cranial nerves are usually affected only when the limbs and trunk are more or less completely paralyzed.

The ordinary form of multiple neuritis, in which there is widespread palsy, can be scarcely mistaken for *tabes dorsalis*, but in the cases in which sensory loss and ataxia are the most prominent features diagnosis may be extremely difficult. Neuritis is, however, distinguished by its more rapid evolution, and by the absence of lightning pains, of sphincter disturbances, optic atrophy and alterations in the pupillary reactions. On the other hand, in *tabes* there is, as a rule, no true paralysis, and when muscular atrophy does occur it is usually limited to one group of muscles, and is rarely symmetrical from the beginning. But the most valuable diagnostic sign is that while in neuritis the nerves and muscles are invariably, in some degree at least, tender to pressure, in *tabes* the pain reaction on squeezing the nerves and on deep pressure on the muscles is almost always diminished and frequently quite absent. The history of infection, an excess of lymphocytes and globulin in the cerebrospinal fluid, and a positive Wassermann reaction are in favor of *tabes*.

The symptoms of *trichiniasis* may resemble those of polyneuritis, but the acute onset with fever and gastro-intestinal symptoms, the œdema about the eyelids, and the swelling and tension of the affected muscles, as well as the character of the pain in them, are fairly characteristic. The eosinophilia and the early affection of the functions of the cranial nerves are also distinctive features.

With *general œdema* there is often a certain degree of flaccid paresis of the limbs, especially of the legs, and loss of the tendon-jerks. In some of these cases there may be a slight peripheral neuritis due to a general toxic state, but the rapidity with which the paresis disappears, and the early return of the tendon-jerks as the œdema subsides, makes it more probable that the paresis is a direct mechanical or nutritional effect of the effusion into the muscles.

*Acute myositis* may be diagnosed from polyneuritis by the swelling of the extremities owing to inflammatory œdema of the subcutaneous tissue and muscles, the rigidity of the muscles, the absence of sensory disturbances, and the erythematous rash which is nearly always present over the affected parts. Finally, in myositis the knee-jerks often persist. A form of paraplegia in old people due to contractures following chronic myositis, which has been described by Lhermitte<sup>1</sup> and others,

<sup>1</sup> *Nouv. Iconographie de la Salpêtrière*, 1906, xix, 251, *Thèse de Paris*, 1907.

may be confused with polyneuritis. But it rarely occurs before the age of seventy-five, and generally only in feeble persons confined to bed. Like neuritis, it may set in with pains, cramps, and weakness of the legs, but it can be easily distinguished by the persistence of the knee-jerks, the normal electrical reactions of the muscles, and the firm contracture of all the muscles which are affected.

**Prognosis.**—The outlook naturally depends on the acuteness and severity of the attack. In the early stages, when the paralysis is spreading rapidly, prognosis must be guarded, as there is always the danger that the affection may involve the cardiac or respiratory apparatus and lead to sudden death. But it is often surprising how patients in whom the diaphragm is paralyzed, or the heart dilated, irregular, and rapid, recover. The disease almost invariably progresses for a time after the poison has been withdrawn, but when its course once becomes stationary there is very little danger of relapse, even though improvement is slow. The chief danger to life results from the complications, especially from tuberculosis; if these can be excluded, and the cardiac and respiratory functions are not disturbed, the outlook as regards life is very favorable and the chances of ultimate complete recovery are excellent. The restoration of function is always slow and even in cases of moderate severity four or six months must elapse before the more severely affected muscles have fully regained their power. The electrical reactions are a valuable guide in estimating the duration of the muscular paresis.

**Treatment.**—The first essential is to discover and remove the cause, or stop its action as soon as possible. In dealing with alcoholic neuritis this is often not easy, especially with female patients who take every possible means to deceive the medical attendant and obtain further supplies of alcohol. Frequently it is necessary, and when possible it is always advisable, to place the patient in a home or hospital under the care of trained and reliable nurses; if he remains in his own home he will probably succeed, even if bedridden, if it is in any way possible, in procuring stimulants. Many authorities recommend that alcohol should not be suddenly stopped, owing to the danger of collapse; but this is rarely to be feared except when the heart is very feeble, and it is certainly inadvisable to continue the administration of a drug which is producing such serious symptoms.

*Rest in bed* is always advisable, except perhaps in the mildest cases; the patient is thus most easily protected from exposure to cold, which may not only have an unfavorable influence on the neuritis, but also make him more liable to pulmonary complications. Complete rest is also a matter of great importance, as movement often seems to intensify the changes in the degenerating nerves, and it certainly increases the pain. In severe cases the use of a water or air bed is generally advisable, as on it the necessary pressure is more uniformly distributed, and the patient does not suffer so much as when his limbs lie on a firmer and ill-adapted surface. Much relief can be given by removing the weight of the bed-clothes by bed-cradles.

There is no specific treatment; it can be, therefore, in the acute stages at least, only symptomatic, and the most urgent and prominent symptom

is usually pain. Often it is necessary to have recourse to the use of drugs; antipyrine and phenacetin are the most effective, and there is rarely any contra-indication to their use; when the patient is restless and sleepless, bromides may be combined with one or other of them. Occasionally the pains are so severe and constant that morphine is necessary, but it is evidently advisable to avoid its use in a person already addicted to a drug habit. Local applications over the chief seats of pain often give considerable relief, especially warm fomentations. Mills recommended the alternate application of very warm and cold water with a large sponge over the limbs, and Pospischill cold packs. It often suffices to keep the limbs wrapped up in a thick layer of cotton wool. Oppenheim has obtained excellent results by diaphoretic treatment; warm baths may give relief if the patient is strong enough to be moved from bed, but this should not be attempted if the heart is feeble. Local hot air baths, which can be used in bed, are sometimes very effective, and if they are not available warm packs may be employed.

Another important aim should be to prevent the development of contractures. The greatest danger is that the feet become fixed in the ankle-drop position, or the hips and knees in semiflexion. From the first, steps should be taken to keep the feet at about right angles to the legs, by the use of a large sand-bag at the foot of the bed, against which the soles of the feet may press, or by a broad, padded board placed vertically across the bed. If the calf muscles are already shortening, their contraction may be overcome by the constant tension of rubber cords stretching from the toes of shoes which the patient wears to a band around the limb at about the level of the knee. In the acute stages the knees are nearly always held semiflexed, but this should never be permitted, as the danger of shortening of the hamstrings is very great.

Of the many drugs, salicylate of soda and iodide of potassium are probably the most frequently used, but there is no evidence that they have a specific effect. Attention is better directed to general tonic treatment; strychnine in fairly large doses is certainly of considerable value when there is any tendency to respiratory embarrassment or cardiac failure. Arsenic should be rarely given, and only in small doses and under constant supervision, as it itself may be a cause of neuritis. Diet needs some attention; it should be as light and nourishing as possible.

During the stationary stage and when improvement is setting in galvanism may be employed, but it should not be adopted in the early stages when tenderness of the muscles is still very great. Large electrodes should be used, and the current should be the weakest that can produce a definite contraction of the muscle fibres and yet is not strong enough to give pain. Massage also may be of considerable service, but it should not be resorted to in the acute stages or while the limbs are still very tender. At first it should be as gentle as possible.

The patient should be encouraged to move the affected limbs as much as possible, when movement does not give him any acute pain, as voluntary movement is undoubtedly a more effective stimulus to nutrition than either galvanism or massage, and voluntary effort probably exerts a favorable influence on regeneration. If contractures have been allowed

to develop, tenotomy may be necessary, but they can be generally overcome by systematic passive movements and extension.

**Acute Infective Polyneuritis.**—The group of cases included under this title has been variously described as rheumatic, infective, or spontaneous neuritis, as it is rarely possible to ascribe the attack to any definite cause; or as febrile polyneuritis, since the disease usually sets in with febrile symptoms. Many cases follow or are attributed to cold, exposure, or overexertion, but it is difficult to see how these factors can directly produce such widespread and progressive symptoms, or the extensive parenchymatous neuritis to which they are due. It is very probable that the polyneuritis is directly due to some bacterial infection, which may not be always of the same nature. Its onset is frequently preceded or accompanied by general malaise, pains in the back or limbs, a rise of temperature to perhaps 104° F., gastro-intestinal symptoms, and loss of appetite. In the severer cases the attack may be ushered in with a rigor; swelling of the spleen, profuse sweating and albuminuria, as well as vomiting and diarrhoea, have been observed. Its infective nature is further supported by the occasional occurrence of small epidemics, for which no definite cause can be discovered.

Generally, as the febrile symptoms are subsiding the patient begins to complain of numbness, tingling, or other subjective sensations in the fingers and toes, and occasionally of sharp or burning pains, but these are rarely so severe as in alcoholic neuritis. Within a short time some loss of muscular power appears, generally in the legs before the arms, but it is more or less symmetrical and greater in the distal than in the proximal segment of the limbs. In rare cases the arms are affected before and more severely than the legs.

The disease is a typical parenchymatous neuritis. Its clinical features are practically identical with those which have been described under alcoholic neuritis, for no set of symptoms is exclusively related to a single cause; but there are a few characteristic points. The *paralysis* is, as a rule, more generalized than in alcoholic neuritis of corresponding severity, the trunk muscles are usually relatively more affected, and even complete bilateral flaccid facial palsy is by no means uncommon. Cases may be seen in which, though the limbs are not very severely affected, the patient is unable to close his eyes, his articulation is indistinct owing to loss of the labial letters, and as he attempts to drink, fluids dribble from the corners of his mouth owing to the palsy of the lips. Not infrequently, too, the palatal and pharyngeal muscles are weakened in some degree, so that swallowing becomes difficult and food may regurgitate through the nose. Laryngeal paresis, producing aphonia, is rarer.

There is also, as a rule, less sensory disturbance than in the alcoholic cases; spontaneous pains may be slight and even occasionally absent, and although the nerves and muscles are always tender to firm pressure, they are rarely as extremely so as in alcoholic paralysis.

This form generally runs a fairly rapid course, often reaching its acme in a week or ten days, and then, after a stationary period in favorable cases, the patient begins to improve. Fatal terminations, which are not infrequent, generally result from paralysis of the respiratory muscles or

from heart failure. Acute cases which die within ten days may so closely resemble Landry's paralysis that distinction is difficult if there is but little sensory change, or if the evolution of the disease has not been observed. There is, however, generally more constitutional disturbance than with the onset of Landry's disease, and the early affection of the cranial nerves in the neuritis is an important point in the diagnosis.

**Recurring Polyneuritis.**—Under this term Mary Sherwood<sup>1</sup> described two cases in 1891, although one of them, in which only the nerves of one arm were affected, certainly does not belong to this class.

These cases are characterized by a marked tendency to repeated attacks of multiple neuritis for which no definite cause can be ascertained; cases in which the successive attacks are due to renewed exposure to some poison, as alcohol or lead, are naturally excluded from this group. H. M. Thomas,<sup>2</sup> who reviewed the subject in 1898, was then able to collect only seven cases, and Hoestermann,<sup>3</sup> who published three original cases, could only add three additional records. The one observed by Thomas is most typical: A man, aged twenty-eight years, otherwise healthy, who did not indulge in alcohol to excess and was apparently not exposed to any poison, developed each June for five years an attack of typical multiple neuritis, from which he slowly recovered in four to six months. Remak and Fletan<sup>4</sup> point out that the disease is often atypical, and that one or more of the cranial nerves have been frequently involved. It is difficult to say whether the repeated attacks are the result of an unusual predisposition, or if the first attack leaves the nerves more liable to a second. Dejerine-Klumpke and Bernhardt have observed recurrent lead palsy apart from renewed exposure to the poison. One of these cases reported by Barnes<sup>5</sup> as "Toxic Degeneration of the Lower Neurones," and that recorded by Williamson<sup>6</sup> under the same title, probably belong to this class.

<sup>1</sup> *Virchows Archiv*, 1891, exxiii, 166.

<sup>2</sup> *Phila. Med. Jour.*, 1898, i, 885.

<sup>3</sup> *Deutsche Zeitschr. f. Nervenheilk.*, 1914, li, 116.

<sup>4</sup> Nothnagel's *Spezielle Pathologie und Therapie*, 1900, Band xi, 3te Teil.

<sup>5</sup> *Brain*, 1902, xxv, 479.

<sup>6</sup> *Ibid.*, 1903, xxvi, 206.

## CHAPTER XV.

### EPILEPSY.

By L. PIERCE CLARK, M.D.

**Synonyms.**—Epilepsy was derived originally from the Greek *epilepsia* (*epi*, upon, and *lepsis*, seizure), a disease of seizures. The word persists with the original significance in the Latin, *epilepsia*, French, *épilepsie*, Italian, *epilessia*, etc. The Romans designated the affection as *morbus commitialis* (the attendant malady) and also as *morbus caducus* (falling sickness); in the latter form it persists in most European languages—German, *fallsucht*, *fallende sucht*; Italian, *mal caduco*; English, falling sickness, etc.

**Definition.**—Epilepsy is a chronic disorder of the central nervous system characterized by more or less frequently recurring convulsive attacks attended by a loss of consciousness. Both phenomena may vary much in duration and intensity.

**Introduction.**—The number of individual elements to be considered in epilepsy is very large, and includes the seat of the disease, which is the entire brain, the motor and psychical phenomena being referable to the cortex. The overexcitable brain and a peculiar psyche may exist without active manifestations (latent epilepsy). Local lesions in the brain, peripheral irritation, may gradually produce the epileptic brain. The seizure types include the major convulsive paroxysms (grand mal), usually complete, but at times incomplete or atypical, and petit mal, or the abortive seizure. In the typical major seizure there should be prodromes (aura), loss of consciousness, convulsions, and a post-paroxysmal state of exhaustion, stupor, and confusion.

While we speak of focal epilepsy or Jacksonian spasms as distinct from genuine epilepsy, being of organic origin, the latest authors tend more and more to break down the wall between. It is still convenient, however, to distinguish between *true*, *genuine* or *idiopathic* epilepsy, and seizures due to actual macroscopic lesions known as *organic* or *focal*.

The *interparoxystic state* and *epileptic mental deterioration*—which supervene surely as a result of the injury inflicted by years of convulsions—are allied subjects which belong to the psychic expressions of the disease. The so-called isolated *psychic equivalent*, the existence of which is disputed by some authors, cannot readily be separated from post-convulsive confusion, and may be considered under the same head. Confusion, with hallucinosis, leads the patient to perform foolish, immoral, and often criminal acts, and forms an important feature of forensic medicine.

In no country is the exact number of epileptics known. It is probably not far from 1 in every 300 to 500 of the general population in the United States. In regard to *sex*, figures show much variation. Gowers finds



women to outnumber men (43 per cent., male, 57 per cent., female; also Morselli, 46 per cent. male, 54 per cent. female). In most reports the males are in excess, either slight or considerable (Binswanger, 62 per cent. male, 38 per cent. female). The ages are no doubt responsible for want of parallelism, as the sex frequently varies with the decades. The *age* at the outbreak of the disease varies. According to Gowers about one-eighth of all cases begin in the first years of life.

As regards deaths in epileptics: The *status epilepticus* is the cause of at least 20 per cent. of the deaths to be directly ascribed to accidents of the disease itself, while 20 per cent. are due to œdema, congestion of the lungs, aspiration pneumonia, and the like. Asphyxia in attacks shows approximately 4 per cent. of the deaths. The epileptic deaths in Hahn's statistics stand in relation to the general deaths as 343 to 541 (63.4 per cent. of the whole deaths). Of deaths not ascribed to epilepsy we note a relatively low incidence of tuberculosis, influenza and lobar pneumonia, cancer, nephritis, cirrhosis of the liver, diabetes, etc., which appears to show that the epileptic either *per se* or as a result of segregation shows a remarkable immunity to certain world plagues. Deaths by means of the heart, vessels and lungs are very numerous.

The *mortality* of epileptics is not much above that of the general population and is much lower than that of the insane as a class. However low the mortality among epileptics is, their longevity as a class is not as great as the average man. The epileptic insane perish nearly ten years earlier than the epileptic not so designated. Mortality increases both with the menace to life from the disease and the increasing age. The menace to life is in part due to accidents. The not very great increase in mortality over sound subjects is to be traced to the number of convulsions and cumulative or progressive character of the disease, and this is offset in part by robustness of constitution. If the dangerous accidents could be eliminated, the result would favorably influence the percentages. This is largely realized in colony life. The mortality of colonized epileptics is naturally much less than in uncontrolled epileptics.

**Etiology.—Predisposing Causes.**—The most striking fact in connection with the hereditary nature of epilepsy is that at least 20 per cent. of epileptics present no evidence of neuropathic ancestry or indeed of any other causation. That epileptic children should be born of neuropathic-degenerative stock, including alcoholics, is almost a foregone conclusion; but in most cases epileptics do not beget epileptics. From a common stock we see develop neuropathy or psychopathy, degeneracy, imbecility, insanity—expressed very largely in antisocial units like paupers, drunkards, vagrants, prostitutes, etc. In other words, an epileptic tends to be born from such stock. But the epileptic may at times descend directly from the epileptic, and in any case, the epileptic brain is not merely a psychopathic brain, but a something added. Literature abounds in statistics of heredity. Moreau noted years ago that one-sixth of his epileptics were of epileptic parentage.

Nothing has as yet superseded Echeverria's statistics. He was able to trace the progeny of 136 married epileptics (one or the other parent) who procreated 553 children. Of this number 78 were epileptics while

185 died of infantile convulsions. This author, by using all available statistics, concluded that 14.10 per cent. of all epileptics have direct epileptic ancestry—a figure doubtless too small, if we reckon some of the victims of infantile convulsions as epileptic. *Familial epilepsy* is not rare. The best examples of this are to be found in myoclonus epilepsy.

Why the ovum is of such constitution as to develop a psychopathic or epileptic brain is, of course, impossible to answer. Hence there has been much speculation as to the part played by conception while intoxicated, and the excessive use of alcohol and other race poisons after conception—upon the supposition that the highest faculties suffer first and chiefly.

The closest equivalent of epilepsy is imbecility. The two constantly occur in relatives or in the same subject. Children begotten at times when a large proportion of a community is presumably intoxicated, rather tend to imbecility than epilepsy, although the latter disease may show some increase. In alcoholism in the parents continental statistics usually show an unfavorable influence upon the ovum; but in some figures relating to England quoted by Turner, children of alcoholics showed no especial tendency to the disease, even the reverse.

The influence of “race poisons”—minerals and toxins of disease, upon the embryo and fetus (without special regard to syphilis), has shown ever that such children are born small and of low vitality, nutrition, etc., but without any especial blight on the brain. That is, these substandard children may reach complete development, but are small and puny and with low vital capacity. The children of victims of saturnism are said to have convulsions.

The question of maternal impressions, *i. e.*, violent shock to the pregnant mother, seems of some minor importance. The women will chiefly be found of neuropathic stock. This origin, however, applies to all forms of psychopathy and degeneracy. Violent mental or mental and physical shock in a gravida may be followed by the birth of a degenerate child, a “black sheep” so-called in a sound stock.

*Birth traumatism*s, in addition to prenatal lesions, are doubtless able to “epilepticize” a brain, especially if the stock is tainted. Molecular changes may exert the same power (Binswanger). Another aspect of epilepsy in the infant is its relationship to simple eclampsia infantum. It is only recently that the labors of pediatricists like Theimich and Birk have shown that there is no actual connection between the two although they may both develop upon the same soil. Infantile convulsions in the first weeks of life are as a rule a sign of incapacity for life. Theimich and Birk followed 53 such cases up to the ninth to twelfth year and not one became epileptic, although not a few showed mental inferiority. Authorities like Redlich and Potpeschnigg have good arguments to the contrary, such as the occurrence of the Chvostek sign in epilepsy, and the not infrequent passage of spasmophilia into epilepsy. In a particular series of cases infantile convulsions led up surely to epilepsy.

When infantile convulsions are carefully analyzed, they will be found to differ notably from an epileptic paroxysm. There is always some reflex irritant and the temperature rises. The cerebrum is involved

very deliberately. A preliminary stage consists either of general motor unrest or stupor with staring eyes and dilated pupils. Convulsions then begin in the facial muscles. Loss of consciousness is a later development. Tonic contractions appear in the lower jaw, larynx, pharynx, and tonic-clonic movements in the extremities.

A degenerative stock is but slightly in evidence in the first six months. There may be only increased reflex excitability. Other authorities see a proved connection between infantile convulsions and nutritive disturbances. Aside from these, certain irritable zones, etc., give rise to reflex convulsions; these include the gums, stomach, urinary organs, and others. In addition to all these causal factors one may invoke an hereditary factor in about one-half of all cases.

It seems reasonable to infer that about one-half of all epileptics owe their disease to antenatal factors, which eventually lead up to the typical epileptic brain. The disposition is there and the causes which bring it out may be mild or severe.

In analyzing the subject of etiology into its original elements, we find that intoxications and infections play a leading rôle. This naturally leads us to the part played by the acute and chronic infectious diseases, with especial reference to tuberculosis, syphilis, alcohol, and all other world plagues. Marie and others lay much stress on such diseases as scarlatina, pertussis and typhoid of childhood as conditions which predispose to epilepsy. Binswanger and others have written at great length on the complete relationship of epilepsy and syphilis. As already stated, very few cases of the former react positively to the Wassermann test. This small number does not include the paretics who as a class are subject to convulsions. There is but a single established type of syphilis-epilepsy, to wit, the Fournier, due to focal syphilitic lesions in the brain. In rare instances of the Fournier type, no gross lesions are present. There may be local spasms due to lesions in one or both hemispheres. The brain then may undergo the special epileptic alteration and general convulsions develop. Recent syphilis may light up epilepsy in the predisposed.

Binswanger, who after infinite pains is admittedly unable to solve the riddle of syphilitic epilepsy, states that the latter expression is merely a collective term for any kind of spasm occurring in syphilis. There are, despite all adverse criticism, two distinct forms of syphilis of the brain: (1) para- or meta- or post-syphilis, and (2) the tertiary gummous syphilis. The former, despite what is said adversely, may occur at any period of syphilis, beginning with the secondary. Paresis and tabes are later developments. The tertiary gummatous syphilis may be placed under the focal or Jacksonian type. The subject of syphilitic epilepsy is at best unsatisfactory to deal with. Why should not an epileptic youth contract syphilis? Why should he not develop paresis? Why should not a syphilitic infant become an epileptic of the ordinary sort when we consider his usual ancestry? But after all, when but 2 or 3 per cent. of epileptics taken at random give the Wassermann reaction, why does not this small coincidence of the two diseases show that the subject is not one of prime importance?

Convulsions of various kinds occur in users of spirits, but in ordinary alcoholism epileptic attacks are not very frequent. It is admitted that neuro- and psychopaths and epileptics usually offer but little resistance toward alcohol. Alcoholic indulgence often causes relapses.

In the convulsive and delirious reception-ward at the Berlin Charité there was a notable association between these conditions. The figures showed that from 36 to 40 per cent. of deliriants were epileptic. In alcoholic mental disturbances there was about 10 per cent. of epilepsy. Fürstner found that nearly one-third of delirium tremens cases were epileptic.

**Trauma, etc.**—Nothnagel has seen a case with positive clean history become a confirmed epileptic after a blow on the head. A somewhat different type exists in which the injury causes fractures of the skull and wounds of the brain, with scar formation. The injury causes both shock and irritation. These stand in some relation to Jacksonian as well as ordinary epilepsy. The relationship of injuries producing simple concussion to epilepsy has caused authorities to suspect that many cases of epilepsy dating from early childhood may have originated in overlooked falls—a favorite view of the laity.

Injuries and other sources of irritation at a greater or less distance from the brain, have been held responsible for a type of epilepsy familiarly known as “reflex epilepsy.” The confusion here is very great; even lesions within the cranium may act peripherally on the cortex. The subject is best studied when cortical foci alone are present. The action upon these of the “summation of irritations” might gradually extend from the foci to the entire cortex until an epileptic brain results. But within late years the distinction between true and partial epilepsy is gradually disappearing. The infracortical, partial and Jacksonian spasms are considered to be of the same nature. From the cortex both local and general convulsions arise; from the infracortical ganglia there arise tonic spasms, shivering or trembling spasms, entirely nonepileptic in character.

In the narrower sense of the term “reflex epilepsy” we refer to epileptic convulsions associated with injuries to peripheral nerves. There may be convulsions of this origin which are by no means epileptic. Merely regional spasms may be produced by severe irritation of the nerves; among these are included tremulous and shivering movements. In such cases there is extension to other muscle groups until the entire body is involved, but these convulsions never extend above the cord, and seldom cross from one side to the other. The cortical symptoms are sensory—vertigo, confusion, and complete loss of consciousness.

That attacks of epilepsy may begin with certain peripheral irritation and continue until the latter is removed is attested by very many cases. That caries of the teeth may be the cause of the paroxysms, which may subside or disappear, is vouched for by Féré. An epileptic who had not had an attack for ten years developed one on the breaking through of a wisdom tooth, and had consecutive attacks. Many similar cases have occurred in the nasopharyngeal and aural regions.

We may make three groups of reflex epilepsy: (1) Where the relationship is obscure; where trauma causes ordinary epilepsy, and where

connection between wounds and spasms is not absolute. (2) Where the patient may have had a strong inborn tendency to epilepsy, and there may be no direct evidence that the injury caused the spasms. (3) Where the relationship between scars and convulsions may be demonstrated at will. Members of the latter class possess the so-called epileptogenous zones, chiefly scars. A convulsion may be caused by pressure on a scar. If we review these epileptogenous zones they appear to consist of two types. In one the zone lies in the distribution of the injured nerve. In the second type there is no apparent connection between the epileptic zone and the injured nerve. The convulsions do not appear to be epileptic, but local and psychogenic.

**Psychic Influences.**—These are well known for their power to precipitate individual attacks, as a rule only the first one. According to Gowers this holds good for 75 per cent. of cases. The emotions concerned are not necessarily those of anger or terror, for they are at times joyful. In such cases, however, the patient will be found neuropathic, probably with a labile vasomotor system. Fright seems to be active at puberty, especially in girls. According to Gowers, there is always an interval between the fright and the fit, but Binswanger finds many exceptions.

Psychic infection—the spread of a case to bystanders—has often been seen. In the Middle Ages, some towns officially regarded epilepsy as contagious. No doubt these were examples of what is now termed affect epilepsy, the number and variety of which are constantly increasing. The subjects are psychopaths, and exhibit seizures which in many cases can be differentiated only with great difficulty from true epilepsy. The seizures are chiefly psychogenic in character and naturally resemble hysteria. In institutions seizures may occur at any friction with officers or other inmates. Fever, great heat and alcohol act like psychic insults.

The so-called epileptiform convulsions due chiefly to poisoning of any kind, whether exogenous or endogenous, are no longer separated radically from epilepsy, because when not mortal in character they are able at times to set up genuine epilepsy; of crises of any kind, fits of laughter, coitus, etc., the same may be said.

Many external factors, such as hypertrophy of the heart, poorly refracting eyes, etc., while not in evidence as causing crises, have much to do with their production. Thus in 61 per cent. of cases of epilepsy there are errors of refraction and other anomalies of vision.<sup>1</sup> Properly fitted lenses often cause improvement and sometimes recovery. The influence of the heart is so marked in certain cases that the condition is termed *cardiac epilepsy*. In such cases cardiac sedatives form a necessary ingredient in the therapy.

Despite all the preceding evidence in regard to the first attack, in a third of the cases submitted to statistics there were absolutely no causal factors discoverable (Lömer).

Special forms of true epilepsy which may be considered from the etiological standpoint are not numerous. We have a few exceptions, as

<sup>1</sup> Schön and Thorey, *Arch. f. Psych.*, vol. xxxix.

*nocturnal* and *menstrual*. The influence of the weather—attacks appear more readily during any change of air pressure—gives us nothing by which to name a class. While mental overwork is hardly enough to determine attacks, it is of undoubted influence as a contributory element. There is no doubt that dreams have determined many first attacks but such instances are more properly classed with so-called affect epilepsy.

In speaking of prodromes we have to consider not only the near but the so-called remote type. This, being volatile and vague, is not often seen; we do not know how often it occurs. This question is, however, of great forensic value. Several days before the paroxysm a distinct psychical and nervous alteration is perceptible. The disposition becomes irritable; small occurrences cause anger, contrariness, and suspicion. The patients feel injured, insulted, suppressed, and yield to impulses of violence. They also feel depressed, ill, uncomfortable, and apprehensive, with a sense of tension or pressure in various localities. Fearful dreams and insomnia are powerful factors in the psychical prodromes. As the hours wear on headache becomes worse, there is confusion, vertigo, and inability to do manual labor. The limbs are tired and heavy, the skin has burning and creeping sensations, and there are nausea, yawning, and hiccough. The part played by the vasomotor system is marked, and may affect nearly all the tissues, notably the mucosæ.

The "epileptic psyche" is fully in evidence in epileptic idiots and degenerates and old epileptics. While it is true that chronic epilepsy is usually complicated with some other mental state, old or new, there is a class in which the brain was evidently intact before the disease developed. Yet with intact brain there were nervous peculiarities. The patient might be termed temperamental in certain directions; he is a neuropathic, exhibiting many of the severe phenomena of this condition. The epileptic psyche is perhaps best seen in children. They present an odd mixture of confusional states, frequently accompanied by excitement and violence. They often vary their states of irritability with cruelty (that shows strong sadistic factors) and dullness and apathy. The rapid and seemingly unaccountable change of temperament and mood is strangely misinterpreted until the epileptic state is fully disclosed by paroxysmal phenomena.

**Pathology.**—This subject is comprised under three heads, viz., gross anatomical changes (*per se*), minute anatomical changes (*per se*), and finds which connect organic with genuine epilepsy (Binswanger).

**Gross Changes.**—These are found in great number and variety in epilepsy but in general are non-specific in character, for they occur also in idiocy and infantile cerebral palsies. They represent congenital errors of development or lesions apparent soon after birth and in earliest infancy. We have to distinguish between simple arrest of development and actual pathological processes. Thus numerous conditions coming under the head of scleroses are really examples of the former (tuberous form, diffuse and circumscribed sclerosis, the so-called stadium verrucosum cortex). Porencephaly, a defect of fetal origin, is occasionally present. Of especial interest is a form of lobar sclerosis limited to Ammon's horn which is so often seen (50 per cent.) in epilepsy. It has been a subject of

debate for decades, and opinions still differ greatly as to its nature and significance. The cornua are firm and anemic to the naked eye. Lobar sclerosis in general is due to the condition of numerous diffuse foci. The whitish appearance distinguishes them from the normal gray matter. They may be extremely shallow (plaques) or of various degrees of depth, smooth or scarlike.

Passing to the other group which are not due to mere arrest of development and which may develop during the fetal and early infantile period, we find chiefly the results of inflammatory processes—encephalitis and meningitis with resulting hemorrhage and softening—conditions which are, in fact, the essential causes of infantile cerebral diplegias; the traumas of birth, which stand also in direct relationship with the latter and which comprise depression of the skull, laceration of the intracranial tissues and their consequences. Much more rarely are encountered finds which are not inflammatory or traumatic, as chronic hydrocephalus, cysts, and tumors. But these which we associate with the newly born or very young child do not differ radically from those dating later in life, which include trauma of all kinds, the results of intracranial inflammations, tumors, granulomata, cysts and in later years arteriosclerosis.

**Minute Changes.**—So constant is the association of gliosis with epilepsy that with some writers the one implies the other. Alzheimer found the condition termed by him marginal gliosis in 40 per cent. of all cases. The excessive proliferation of the glia, however, is clearly secondary to changes in the neurones. Some of the nerve fibres and cells disappear altogether. Such changes have been noted in comparatively recent cases and in status epilepticus. Alzheimer (confirmed by Clark and Prout) sums up the combined changes briefly as follows: proliferation of blood-vessels, large amoeba glia cells, division of nuclei in glia cells, regressive changes in ganglion cells, disintegration of axis cylinders. In a much larger percentage of cases Alzheimer evidently found no marginal gliosis, and in fact nothing to throw light on the disease. Sclerosis of Ammon's horn, so often present in genuine epilepsy as to be almost typical, he appears to regard as secondary and degenerative. Finally in a very small fraction of cases the findings consisted of atrophy of a few convolutions with marked disappearance of nervous elements and some increase of glia. It should be remarked that the (secondary) gliosis of epilepsy is in a sense specific, and quite unlike any other type of gliosis. Recent studies of the finer anatomy of sclerosis of Ammon's horn show hypergliomatosis associated with actual disappearance of the nerve cells, most strikingly seen in the pyramid cells. The two were complementary, varying directly with each other.

**Relations Between Structural Changes and Epilepsy.**—This subject has been foreshadowed in preceding paragraphs and is discussed in other sections. It can hardly be dealt with save in a cursory manner. The original belief that Ammon's horn is a powerful motor centre having been dispelled, lesions of this body can no longer be regarded as causes of epilepsy. Marginal gliosis is looked upon by Alzheimer as standing in direct relation to mental deterioration and imbecility, which is one of the most striking components of epilepsy. Focal lesions in very young

children are believed to have the same force as congenital defects, since they lead to arrest of development. Such lesions may readily associate themselves with typical genuine epilepsy. It is impossible to detect any relationship between these lesions and any particular type of true epilepsy. Marginal sclerosis is almost never associated with sclerosis of Ammon's horn. In theory porencephaly should be an ideal anatomical condition for association with epilepsy, but few cases of this association occur. The epilepsy which accompanies infantile cerebral palsies is genuine. Jacksonian epilepsy does not as a rule belong to autopsy finds.

**Symptoms.**—The *aura* is the first portion of the epileptic seizure, but does not appear in over one-third of all cases. It is frequently purely sensorial and remembered by the sufferer, which shows that it antedates the practically constant loss of consciousness and fall. The more rapid and explosive the attack, the less the opportunity for an aura. In such cases, on the other hand, the remote prodromes already described are relatively common, acting as an aura equivalent. Gowers makes no less than seven types. These, however, may be condensed to five:

1. *Psychic Auras*. These are emotional or intellectual, the former characterized by fear or anxiety, euphoria, joy, etc., appearing suddenly or after painful expectation. In the latter all the mental faculties or the memory alone may be suppressed. In rare cases of absence of convulsions petit mal is closely simulated.

2. The *sensorial auras* affect the eye and ear and rarely the other special senses. The eye may see as if through a fog or behold formless colors, sparks, scintillating scotomas, etc.; there may be hallucinations of all kinds. The ear may hear loud noises, reports, musical sounds, and there may be otic hallucinations.

3. *Auras of bad smells and tastes*, disconnected from the presence of sapid bodies, occasionally occur.

4. *Sensible auras* differ from sensorial in that they affect common sensibility. They appear as paresthesias or actual pain. Occurring in the toes, the sensation may march up the limb.

5. The *motor aura* is varied, less often seen and is related chiefly to focal lesions of the cortex. It consists of convulsions, locomotor movements, and automatism. The cloni are mere twitchings of muscle groups. Tonic spasms may be bilateral, affecting the so-called "spastic" muscles and following organic brain lesions. They may be ranked under partial epilepsy. The locomotive aura is rare *per se* but not uncommon in nocturnal attacks. They may be unilateral, bilateral, or simply confined to one extremity. The automatic aura appears to be voluntary, as in all automatism. Thus the patient may seem to be trying to abort an attack. The automatism is psychic in origin. The patient is really conscious, and the movements may be recollection pictures. The cursive aura, which is not to be confounded with *epilepsia procursiva*, represents automatic locomotion a few steps forward, before the fall and loss of consciousness occur. The motor aura may take the shape of an inhibitory paralysis. It may also begin with sneezing, yawning, coughing, etc.; or with pains and paresthesia in the abdomen and pelvis.

The *vasomotor* aura is relatively common both as a local or general



angiospasm which begins suddenly. Angioparesis also occurs. Local angiospasm is expressed by pallor, coolness, and sensory disturbances which occur chiefly in the digits. Angioparalytic lesions are seen as blotches, redness of the face, neck and trunk, hemorrhages, etc. When the vasomotor aura is general there is usually a chill, pallor, and burning sensations everywhere, with increased excretion of saliva, tears, and sweat. Such vasomotor phenomena may occur in epileptics independent of seizures, for vasomotor lability is more or less characteristic of the disease.

A single type of aura is not as a rule constant for the patient, who may show a difference with each convulsion. In reflex epilepsy it may be possible to abort the seizure. It is evident that there is no essential difference between certain auras and petit mal.

**Seizure Types.**—Following immediately upon the aura when one is present we see two essential phenomena of ordinary major epilepsy, namely, loss of consciousness and falling to the ground. The patient falls prone in an instant, with all his weight, often on his face. Being unconscious he feels no pain. (There are rare exceptions to the rule of unconsciousness, as shown in a series of my cases in which though the attacks were of a severe grand mal character the individuals were fully conscious throughout the attacks. Most frequently, however, the tonic clonic spasms in such grand mal attacks have not an instantaneous order of muscular involvement in all parts of the body at the same time.<sup>1</sup>) The number and variety of injuries from striking the ground or floor are legion.

The *convulsive* stage is the most striking and characteristic phase. With the patient on the ground or floor, and perhaps in part before he lands on the latter, all the voluntary muscles are thrown in a tonic spasm. The muscles are chiefly synchronously involved, not *seriatim*. The epileptic cry, which occurs only in about one-half of cases, is bound up with the tonic stage but its mechanism is obscure. Since it may precede this stage it cannot be due inevitably to forced expulsion of air. In *tonic* spasms the head is usually thrown back; eyes widely open, staring, and expressionless; face muscles all tense, the jaws locked together; the trunk rigid and immobile, in opisthotonos, or more rarely emprosthotonos; the arms in tetanic extension spasm, rotated inward; fingers enclosing thumb, made into a fist. The lower extremities are likewise in forced extension and inward rotation, thigh abducted, toes spread out and extended or maximally flexed. The cycle of tonic spasms lasts as a rule, and upon an average, about thirty seconds. Relaxation of muscles is not synchronous—as a rule the lower extremities first, trunk next, and head and neck last.

While the tonic phase shows some order, the next or *clonic* phase is irregular, planless and tending to begin in the lower extremities as the tonic spasm wears off. The limbs are flexed and extended in the clonic phase. The variety of clonic spasms is such that they can hardly be described. Then often succeeds simple flexion and extension, inward and outward rotation, etc., the limbs are hurled in every direction, the movements suggesting blows, kicks, stamping, treading, etc. The head

<sup>1</sup> See "Conscious Epilepsy," L. Pierce Clark, *Am. Jour. Insan.*, 1909, lxvi, No. 2.

and trunk are also hurled about and the latter is often made to rotate back and forth. The cloni of the face are the most shocking. In addition to the deformity, these movements cause destructive acts, notably the biting of the tongue. The foaming lips result because the cloni of the chewing muscles churn the increased saliva into a frothy mass. The stomach and intestines fill with gas, followed by much belching. Occasionally there are seminal ejaculations, and involuntary expulsion of urine and feces may occur. A stream of urine may be thrown a distance of 5 to 10 feet at the onset of a severe attack.

The duration of the clonic phase is considerably longer than the tonic phase—from one-half to three minutes. The classical attack is usually bilateral, one-half of the body being involved directly after the other. In atypical and rudimentary attacks fewer muscles are involved. In typical cases we find those violent cloni which cause severe injuries,—fractures and luxations. Cloni are for the most part continuous; exceptionally we see relaxation, followed once more by cloni. The two kinds of spasm overlap in certain seizures.

**Psychic Epilepsy.**—The most striking symptom of this is an alteration of consciousness, which is termed dreamlike. The very mildest phases, which resemble somewhat *petit mal*, are characterized by a brief confusional state, accompanied by a few automatic activities. Cases differ extremely and it is difficult to describe psychic epilepsy in brief, straightforward text. Thus a subject may suddenly run out of doors, return at once, and perform any number of purposeless, automatic acts, some of which may injure himself and others; while acts simple in themselves may lead to arson, suicide, and homicide. A strong sexual bias is often present. The emotional sphere does not enter into these acts, which resemble those of the intellectual aura, or intellectual *petit mal*.

Psychic attacks as just described are really relatively rare in occurrence. Their apparent frequency in many medical reports is due largely to insufficient analysis of such seizure symptoms. The more closely psychic epilepsy is studied the more one finds that the attacks are really preceded by a very slight local spasm of muscles in some one part of the body perhaps, which because of its very transitory and casual nature eludes observation, thus forcing many instances of psychic epilepsy into the medical type of *petit mal* followed by a prolonged train of obvious mental symptoms which characterize the many phases of automatism. So excellent an authority as Hughlings Jackson believed that no form of epileptic attacks occurred without some muscular spasm, however slight.

**Epileptic Psychic Alteration.**—Statements as to the number of epileptics who retain their psychical integrity do not exist. Those accessible to us are institutional cases in which the often associated idiocy shows the frequency of an intellectual defect. With advancing experience we now believe that in many chronic epileptics the mind is intact. By further knowledge of their personalities, various features are seen which point to epilepsy—a certain pedantry and “matter of factness” in all things, extravagance in speech and writing, a distinct egotism or hypochondria, over-appreciation of self, exaggerated sensitiveness—but nothing which points to mental failure. In patients who have attacks years apart we

see no affect phenomena of a chronic character which suggest the epileptic alteration, nor do we see any characteristic defect of intelligence. At times there is a transitory psychic alteration, in relation to affect life (irritability, alterations in temper, outbursts of anger).

The patients who are most in danger of mental degeneration are (1) those who become epileptic at or before puberty; (2) those who have serial attacks, and (3) those who have had numerous abortive attacks.

Of very great interest and importance is the intellectual foundation—the men of great intellect or high spirits who throw off the fits and their results, both readily and without injury. A few world celebrities belong here. Aside from these there are many parallel cases.

In elderly people petty causes seem to be sufficient to cause senile epilepsy, which, with ordinary senile involution already present, rapidly precipitates the latter. There is no general rule applicable to these cases, for I have seen a case of an old man who at the age of sixty-two began to have rapid epileptic attacks. At the age of sixty-eight, after six years of epilepsy he showed no intellectual defects.

The psychic degeneration of epileptics possesses certain characteristic traits, which certainly depend upon the underlying disease and affect disturbances due to the attacks. The intellectual weariness and exhaustion symptoms, which at first follow individual attacks, become protracted in the course of time. Finally a period for recuperation becomes no longer possible. The patients are seen to gradually lose the capacity to receive new impressions. The memory pictures of the newly happened become more scanty and full of gaps. Forgetfulness extends more and more toward the past. We see complete oblivion of certain experiences while for others memory shows no depreciation. The greater the difficulty in receiving impressions the less readily are they reproduced; so that the trend of thought becomes monotonous, while speech expression is awkward and dull. Next comes epileptic dementia, characterized chiefly by attacks of anger. The gracious patient bursts into anger and attacks those about him. Sexual extravagances are common.

**Post Paroxysmal Psychosis (Temporary).**—The transitory psychosis which follows a motor paroxysm is comparable with psychic equivalents, and pre-paroxysmal phenomena (remote prodromes). There appears to be in certain cases a single continuous disturbance of consciousness. But a third of epileptic major seizures are known to have had auras, while the majority have a post-convulsive stuporous stage. One may designate four fairly distinct post-paroxysmal conditions in epilepsy:

1. The more common stuporous state characterized by complete inactivity of mental and physical performances, lasting an hour or so.

2. Attacks followed by a more or less orderly and systematic automatism usually lasting a few minutes to several hours.

3. A fairly systematized series of hallucinations, dream-like in formation.

4. Severe and prolonged states of excitement with pronounced incoherence and violent impulsive activities which may last for several days. These elemental types are frequently combined in many ways. A protracted, stuporous condition, which may last for days and weeks,

is more common as post-paroxysmal state than as psychic equivalent. The relationship between post-epileptic psychoses and attacks is intelligible; the more frequent the latter (until they become serial) the more severe the post-paroxysmal psychoses. The shorter the intermission between individual attacks, the more frequently will the condition be distinguished by a protracted, confused or impulsive, excitable condition.

In a second rare group of cases the transitory psychological disturbance follows individual fully or partially developed convulsions, but at a protracted interval. This sort of post-epileptic psychosis is often of toxic origin; for example, after alcoholic indulgence a number of hours may elapse before the attack sets in. The same is true of an affect and a subsequent attack.

The worst species of post-paroxysmic psychoses are those which follow abortive attacks. It is not easy to state whether the latter are psychic epileptic equivalents with pronounced aura, or simple motor auras, or brief isolated abortive attacks of true epilepsy, or some other known manifestation; whatever the type, the paroxysm is an individualized, brief, self-limited process, be it an attack of vertigo, a brief lapse of consciousness, or a period of violence. Some transitory psychoses do not last over two or three minutes, while the more protracted may last for hours and days, even weeks. These cases of mental cloudiness of relatively brief duration require to be very carefully analyzed in order to fix their status. Let us take cases of "absence." The entire duration of a special case is but one minute with retrograde amnesia.

In simple feeble-mindedness, there is spasmophilia of high degree. But mental defectives after a certain interval of freedom from epilepsy may develop typical attacks of the latter from trifling causes—slight use of alcohol, of certain drugs, fever, or psychic trauma. The spasmophilia or tendency to convulsions in the feeble-minded has been ascribed to infantilism. A spasmophilic imbecile may later develop epilepsy from trivial causes. Difficult as it is to understand the relationship between epilepsy and idiocy in these cases, it is far more difficult to explain cases in which the child was born normal and developed epilepsy in late childhood or puberty. At this very period the intellectual growth is either halted or actually retrogrades.

**Petit Mal.**—A generation ago, medical text-books spoke of petit mal as if it were little more than a mere "nodding" or "losing oneself," which differed from the ordinary form only through the fact that speech, work or other activity was resumed just at the point where the seizure caused the inhibition. Amnesia was present, the patient having no recollection of what had happened. Another characteristic point is that the features instead of being flaccid, are either made tense like a mask or undergo grimaces, chiefly of the ocular and buccal muscles. The condition as a whole became recognized as a form of epileptic seizure, not less deadly in the long run than the severe convulsive attacks; and with this discovery it became known as *petit mal*.

From another viewpoint petit mal maintains its independent character because the simple movements of the facial muscles may be greatly expanded, affecting muscular groups here and there. It is as if when the

inhibition is not so powerful as to cause loss of consciousness, certain motor equivalents set in. The presence or absence of an aura in petit mal is of much significance. In theory petit mal is one form of an abortive grand mal attack. They comprise attacks which consist only of an aura, which are extremely rare and are not followed by a motor discharge, although there are auras which consist solely of certain motor manifestations. Next comes a form of petit mal, in which there is a brief loss of consciousness and at times motor phenomena; finally, with loss of consciousness and face to the ground, we still see irregular and imperfect manifestations of a major convulsion. It is not strange that situated in the midst of abortive attacks petit mal shades into other manifestations. The space given by Gowers and Binswanger to all these phenomena and their mutual relations shows the complexity of this subject.

The subjects of petit mal as a rule remember certain phenomena, chiefly of motor character, which represent auras. In Gower's analysis of 155 cases 45 showed nothing whatever of prodromes. There were not even the premonitory feelings of swoon or drowsiness. In 25 cases there was preliminary vertigo; in 17 shock or tremor of the limbs, and again 17 of eye prodromes—either excessive sensibility or loss of sight. Other elements include mental states (sudden phobias); a unilateral peripheric sensation or twitchings; epigastric sensibility; sudden tremor—and so on down to single causes with but one case to illustrate them.

All these remembered experiences tend to show the great part played by auras in petit mal. The most common feature is vertigo or a feeling of lost equilibrium which does not quite amount to vertigo. About 20 per cent. belong here. But this number can be increased if we include cases which begin as half flexion of certain muscles. As petit mal was first known to the French as epileptic vertigo we can appreciate the value of this connection. Gowers has on several occasions heard the epileptic cry with petit mal, although in grand mal it is usually associated with the fall. The simple loss of consciousness of typical petit mal, with resulting amnesia, represents but a certain number of these cases; others have retention of consciousness. This is likely to be noted in very brief attacks and is usually the result of some physical experience.

It is thus evident that any brief seizure, lasting for a few seconds up to half a minute or more is an example of petit mal; an abortive or otherwise mitigated equivalent of the major attacks. Petit mal stands in near relationship with auras, especially a motor aura. The forms are very numerous—a brief vision or hallucination, leading to defensive motions; twitching of the eye muscles, or of those of one side of the face, consciousness being retained; isolated vertigo may belong here. In a single chronic epileptic, one may see every form of seizure—monospasms with or without loss of consciousness, rudimentary attacks, typical complete major attacks and status epilepticus. A point of importance, according to Gowers, is the great tendency of association of petit mal with vasomotor aura.

**Incomplete, Rudimentary Attack.**—This rare form is known as the apoplectic, because of the clinical parallelism. It is thought to be a

severe form of tonic crisis, which hampers breathing and the cerebral circulation. The post-paroxysmal state suggests an apoplectic stroke.

Paradoxical types occur in isolated cases in which motor auras are closely paralleled. Their culmination is seen in *epilepsia procursiva*. Its locomotor component may be due to the activity of the infra-cortical ganglia: A man, aged twenty-six years, had had seizures of a peculiar character for fifteen years. The first evidence of the attack was the cry but instead of falling he ran forward at great speed, unconsciously paying no attention to the small objects in his way. In a few seconds he stood still, becoming conscious and at the same time very red-faced. He had amnesia for facts but recalled some dream-like image of his experience. A second seizure followed, in which the patient fell but rolled himself over the floor, crying out continually for about twelve seconds. Conditions resembling this have also been described (*epilepsia rotatoria*).<sup>1</sup>

**Phenomena Which Accompany or Succeed Immediately Upon Major Crises.**—Many authors devote much space to the *vasomotor disturbances* during a seizure; they are indeed next to the convulsions the most essential component of the latter. The vasomotor attack is short and quickly replaced by exhaustion, the face becoming cyanotic. This phenomenon, however, may also be explained by compression of the veins of the neck by the rigid neck muscles. The picture shown by the face in severe cases is that of strangulation. With vasomotor spasm there is naturally a sudden and marked rise of blood pressure. As a result, minute hemorrhages often occur, and since these might persist for days often supply evidence of an attack. These occur in the skin over the mastoid process as fine punctiform hemorrhages. Bleeding may occur from nearly all of the mucosæ, but profuse hemorrhages are rare. The very wide divergence of the symptoms of this type overthrows any theory that epilepsy is *per se* a vasomotor affection, while still showing that this symptom plays an important part.

In regard to the pupillary phenomena during and after a seizure there is no complete uniformity, save that dilatation begins with the tonic stage. There is no response to light; after the seizure the pupils usually become normal.

In the *fully developed atypical attacks* the remote prodromes and aura show no peculiarities in comparison with grand mal in general. The differences begin in the loss of consciousness and the order of the motor movements. As a rule the tonic spasms are complete before the slowly progressive inhibitory discharge of the cortex has led to a complete elimination of the psychic functions. This slow onset, aided perhaps by the warning of an aura, gives the patient time to seat himself or lie down on the floor. He often has good recollection of the tonic stage but then loses consciousness. In this form of attack the tonic stage does not develop instantaneously throughout the entire musculature but may begin on one or the other side of the body or in groups of muscles of both sides, and may simulate for the moment coördinate movements.

<sup>1</sup> See "Status Epilepticus," Clark and Prout, *Am. Jour. Insanity*, lx and lxi; also "A Clinical Contribution to the Irregular and Unusual Forms of Status Epilepticus," Clark, *Am. Jour. Insan.*, 1913, lxx, No. 2.

The variety of these irregular tonic spasms is infinite. No absolute borderline can be erected in these cases between tonic and clonic spasm, for at the close there is still some tonus left in the muscles. The post-paroxysmal stupor in such cases is likely to be attended by psychic excitement, but is usually short and followed by deep sleep. From certain facts one may believe that such attacks occur only on a hereditary-degenerative base and that the epilepsy represents but part of it.

The so-called *epileptoid conditions of Griesinger* may be noted here. Most of these may be accounted for under other heads but a few remain to which the title may be given. We designate by the term epileptoid only those recurring paroxysms of disease-phenomena, non-convulsive, with or without consciousness, which have been seen in connection with undoubted epileptic insults, and also appear independently as a sole, unique sign of a discharge. There belong under this head secretory, trophoneurotic, and vasomotor disturbance. Thus a patient may or may not have vertigo at first; but there is a seizure type, in which with no apparent cause a local hyperidrosis appears. Attacks of oedematous swelling may occur as an epileptoid condition, just as in hysteria. This swelling is painful and tender. A third form is sudden darkening of the integument in certain localities of the body. Attacks of urticaria have also acted as epileptoids.

**Exhaustive or Stuporous Stage.**—At the close of a grand mal seizure, the patient sighs deeply and assumes an attitude of extreme prostration and deep slumber. This lasts but a few minutes, when the patient wakes, sometimes suddenly, and makes immediate efforts at orientation. He may jump up and look about, but as a rule his exertions are passive, as if he were trying to collect his thoughts. In a short time, however, the state of exhaustion again overcomes him and he settles down to a condition of sleep or stupor, or at times a sort of delirium, in which dream hallucinations may or may not cause motor activities of various kinds. Various disturbances of speech are especially known. This delirium is really a part of the epileptic psychopathology. The post-convulsive sleep may last from a quarter of an hour to several hours, and almost always there is complete amnesia. The patient is left in a high degree of fatigue, incapable of mental activity, and is sad, irritable and hypochondriacal. He has pains in the muscles and joints, nausea, vomiting, and anorexia. Many subjects feel themselves better off for having had the fit.

The local forms of exhaustion are numerous and not necessarily of much importance. As in other convulsive affections, the muscles may be in a state of paresis. This is also seen in Jacksonian epilepsy. In a few individuals repeated exhaustion of the same muscles leads to an increasing disability. The subject of cortical deterioration, muscular exhaustion after prolonged seizures, and deterioration of the function of the muscles is a complex one. Such lesions as concentric narrowing of the visual field, disturbances of color vision, deafness, etc., exist for a short time as exhaustive manifestations. But little is known of anosmia and ageusia. Much also has been written concerning post-paroxysmal analgesia and anesthesia, together with other disturbances of sensation. These have no diagnostic value and occur without any regularity.

**Rationale of Major Seizures.**—We know the phenomena of seizures from the clinical standpoint, and we know something of the nature of the epileptic brain; but back of all such knowledge there must be a theory or a rationale to account for the why and wherefore of the attack. We have partial ideas of the mechanism of the interruption of a crisis, so that numerous abortive clinical forms occur and they aid us in establishing a theory of the complete major crisis; but it is the latter in its entire cycle which we wish to consider. We also are interested in knowing whether true convulsions can be produced artificially.

We know that the entire brain must be altered in some manner for its owner to become epileptic. We have no word to express this state, because alteration is a vague term, and applies equally to the mental changes which are due directly to the great number of seizures. There should be some term corresponding to spasmophilic to describe the sensitized brain of the epileptic. Many authors contend more or less successfully that the original irritability of the brain differs in no wise from that of extreme cases of neurasthenia (including psychasthenia). Some phenomena predominate in one state, others in the other, but there is no line of demarcation. Other authors, including myself, state that the epileptic predisposition is something altogether peculiar.<sup>1</sup> We are inclined to lean to a dualistic viewpoint, for if an epileptic brain is only a psychasthenic brain, it will be impossible to distinguish radically between true and affect epilepsy. The change in the entire brain manifests itself in cortical disturbances merely because the cortex is the most highly developed portion. All manifestations of general seizures are linked with the cortex.

We cannot take it for granted that the general irritability of the brain is of much help to us in explaining a seizure. Mere irritability must be a factor in all psychoneuroses. We have to account for the sudden character of the seizure and especially for its periodical return. There is a suggestion of petty insults which very slightly irritate the brain, but are not at first manifested. As these insults slowly increase, a period comes in which the brain is comparable to a strongly charged cell. A state of saturation is impossible. Finally a so-called explosion or discharge occurs. In this respect the affection is quite parallel with migraine or any other periodically occurring neurosis. This explosion, at least in so far as it represents migraine, was termed many years ago a "brain storm." The typical epileptic brain, now clear, begins the cycle anew. We cannot, of course, claim that the discharge is complete when frequent or serial attacks make up the disease.

This overloading or discharging causes two types of manifestation, best styled respectively as *inhibitory* and *exciting*. The first inhibit the activities of numerous functions, while the second cause an excess of function. Even the stage of post-paroxysmal exhaustion is not unphysiological; general convulsions cause general exhaustion, local convulsions cause local exhaustion. The first epileptic attack differs from the subsequent ones in being an affect manifestation, due usually to fright.

<sup>1</sup> Clark, *Am. Jour. Med. Sc.*, 1914, cxlviii, 729.



The typical convulsions proceed not only from the cortex but from the infracortical centres, including the large basic ganglia, quadrigemina, and medulla. Certain mild attacks (incomplete) probably concern the cortex alone. It is assumed that the discharge, whether strong or weak, always begins in the cortex. Those convulsions of cortical origin which follow organic disease, injuries, etc., are not at first epileptic but doubtless readily become so.

**Experimental Seizures.**—The subject has been summed up as follows: When a circumscribed area of the motor-excitabile cortex is subjected to a mild electric current, clonic twitchings of the corresponding muscles appear. Nearby muscles also become involved through superficial extension of the irritation in the cortex. After the electrode is withdrawn, the spread of the irritation may reach the cortex of the opposite lobe and cause convulsions therein. In certain animals, and with very strong currents, we may also see tonic spasms and automatic activity (locomotion), evidently from projection to the infracortical motor centres. These experiments show analogy with human epilepsy, and help us to understand some of the rationale, both of general and local attacks.

In man himself there has been much experiment upon epileptics, with particular regard to so-called reflex epilepsy. Experiment may be justified in cranial surgery, in order to aid in the localization of certain motor areas, and there have been many operations performed in the hope of curing organic epilepsy, some of which have experimental significance. But no results obtained can contradict what we already know about epilepsy. An epileptic is not immune to other forms of convulsions such as do not depend upon his peculiar brain texture. He has been known to simulate his own attacks, may have hysterical and psychasthenic crises—in a word, he may have any one of the old so-called epileptiform convulsions. The results of experiment, however they may show the presence of a mechanism also visible in epilepsy, must not be made co-equal with epilepsy as a disease.

*Status epilepticus* is not recognized as a special type of seizure, for it differs only in degree from serial attacks. The latter, if anything, marks any innate peculiarity of seizure. This serial seizure occurs as a rule after prolonged freedom from attacks, whether spontaneous or due to bromides. Another peculiarity of serial attacks is that we find them most frequently after abortive and rudimentary attacks, or the type of the individual seizures may be a picture of all the forms, including grand mal. The immediate consequence of the serial seizures is that the subject has little time to recover from the usual exhaustion and stupor following a single attack—hence when the series ends he finds himself often in a state of prostration and stupor which may last for months.

Status epilepticus instead of resulting necessarily from intensive serial attacks, may set in spontaneously without apparent cause. For a short time individual attacks may be seen but it is not long before we have to deal with one continuous convulsion. In twenty-four hours the subject goes through the equivalent of from 10 to 30 or more separate attacks. The amount of muscular activity, as in the case of tetanus, may send the temperature up to 42° C. or more. A constantly ascending

temperature ends fatally and the temperature continues to increase after death. It can be seen that in status epilepticus seizures of all kinds may succeed one another. The blood pressure is high, the face cyanotic and the subject perspires freely. The condition may simulate rabies in several ways, necessarily in difficult deglutition. The convulsive period lasts from one to three days.

This prolonged convulsive stage is followed by a prolonged period of exhaustion or stupor. Various acts and movements made during this period show that the condition is akin to busy, muttering delirium. The face is very expressive and excited, the gaze wanders, patients toss on the bed and even leave the latter. All the while they mumble parts of words. The patient soon presents a typhoid picture and dies of œdema of the lungs, heart-failure or decubitus.

In patients who are destined to recover we see profound exhaustion, which may last for days. We have here to watch the temperature and should this begin to decline rapidly the patient will recover, if he does not develop the excited hallucinatory period. Recovery may occur under the most unfavorable conditions, a prolonged convulsive state (twelve days) followed by days of excitable delirium, etc., and fever.

Older writers spoke of fragmentary or serial attacks which never led to status epilepticus. Thus 2500 attacks have been experienced in a month by a boy of fifteen (Delasiauve). It is not impossible that these were really psychogenic fits.

**Nocturnal Attacks.**—All are familiar with day attacks, but nocturnal seizures may occur and pass undetected for years. It seems at present that the nocturnal attack is much more common than the diurnal, and that they may precede the latter, perhaps for years. Diurnal attacks, in the belief of some, do not develop until the disease is deep-rooted. Thus nocturnal attacks at puberty may not be discovered until marriage or until a period of military service is required.

**Miscellaneous Seizures.**—An epileptic may exhibit other than epileptic paroxysms, which are best termed *affect epilepsy*. Bratz and numerous others have described this form of epilepsy under various terms. The subjects comprise degenerates, young criminals, etc. Affect influences cause serial attacks, with no tendency to status epilepticus or epileptic deterioration. Such patients have until very recently been interned in epileptic asylums as genuine epileptics. They profit nothing from the hygienic treatment of epilepsy and are never free from pallor. A labile psyche appears to furnish a foundation. Vasomotor lability and spasmodophilia for the most part co-exist. Not only sharp affects, but chronic depression, moroseness, etc., furnish the substratum for these attacks.

**Hysteroid Seizures.**—The new field of affect epilepsy has cut deeply into the old material of hysteria and hystero-epilepsy. Some authorities insist that hysterical and affect crises are one and the same. As in epilepsy, "hystero-epilepsy" occurs on a degenerate soil. The manifestations of the latter are very numerous, and an epileptoid convulsion is but one of them. Oppenheim's psychasthenic convulsions, which are usually ranked under affect epilepsy, are placed as a "third affection" or "bridge" between hysteria and epilepsy, and certainly differ in some ways from

Bratz's syndrome. The latter exhibits a large number of serial fits at long intervals, while Oppenheim's disease shows but one or two attacks in a lifetime. True hystero-epilepsy can only be a distinct combination of both diseases in one subject, each pursuing its own course.

**Psychical Anomalies of Epileptics.**—The psychical phenomena are very manifold. There are two principal forms, transitory and permanent or chronic. These stand in no absolute relationship to each other. First there is the association of idiocy and epilepsy. Statistics show that but one-fourth to one-fifth of all epileptics have a normal psyche. In severe child epilepsy, the convulsions produce an arrest of development, depending somewhat on the age, and but few escape idiocy.

Epileptics are unable to absorb and retain impressions because of dulled perception. The prolonged bad temper has been ascribed to the fact that the results of scanty perception are offset by a much longer period of sinking in of impressions than is the case with the normals. There may be a direct connection between dulled perception and disordered comprehension. Perceptions are often even at the outset falsified by deception of the special senses. Hallucinations and psychic auras before convulsions may be cited first, but in the interval and even in the post-paroxysmal stupor we see the same phenomena and, in the interval state, must often regard them as equivalents. Hallucinations lead up to the dream state and mental confusion or so-called twilight state.

The paranoid states which are seen in combination with epileptic attacks are chiefly manifested by hallucinations. These states are regarded by some as equivalents. Ziehen describes a chronic epileptic hallucinatory paranoia.

Consciousness is the last and highest step of the process of perception. Hence a change in consciousness may mark the very essence of the disease. In severe epileptic seizures consciousness is lost entirely. The epileptic sequence is as follows: dulled perception, and resulting depression of comprehension; spurious perception hallucinations; interruption of perception; loss of consciousness. In twilight states, amnesia is not necessarily present; and in *wanderlust* consciousness is now known to be partly retained in certain cases—imperfect amnesia.

In measuring the degree of intelligence in average men we find in the epileptic a constant diminution of the various faculties. The typical epileptic disturbance of thought is seen in the hazy mental state. Association of ideas is difficult. Inhibition and incoherence are the chief tokens. There is distinct dissociation in the course of thought. The subject performs unexpected acts. Psychical continuity is interrupted. Disturbance of affect life is common in the course of epilepsy. Excitability in dealings with others becomes marked. The least possible offense, real or imagined, causes an outburst of rage. This is especially the case if a little alcohol has been taken. The same outbreaks may occur in stupor, after an attack; others may be regarded as equivalents.

The *disposition* of the epileptic is manifold. He may be depressed, gloomy, preoccupied. This combination of hypochondria and melancholia may last for hours and days. Since they recur periodically they are called equivalents by Kraepelin. They come on without cause,

suddenly, as a general melancholia with its self-reproach and suicidal tendency. Numerous somatic accompaniments appear to be of vaso-motor origin. Intensive variations in disposition also occur in connection with the convulsive seizure. The hypochondro-melancholic behavior may occur before the paroxysm and also after recovery from the latter. The opposite picture, that of mania, is rare. We sometimes see in the post-paroxysmal period a combination of euphoria, increased self-conceit, etc. The psychic aura is at times accompanied by a most intense feeling of happiness or good fortune.

An epileptic is a readily excitable, sensitive man, whom the slightest occasion may cause to explode into violent acts. As a rule he presents a gloomy, closed-in nature. He is suspicious, jealous and vindictive toward others, and is but little interested in the outer world, is dull, indifferent, idle, and performs his duties with difficulty. His makeup in the presence of others is that of an obstinate pedant. His behavior is without form. The smallest trifles he regards as serious, and his opinion cannot be changed. He is notable for piety; is a typical hypocrite, egoist, self-satisfied, and even has marked family pride.

We finally find disturbances in the field of the *will* and *behavior*. A depressed will may be regarded as a psychomotor inhibition, which often becomes a habit. The reaction time is greatly retarded. Inhibition plays a large rôle in the epileptic mood. Extreme disturbances of behavior include arson, theft, etc., occurring in the twilight-state.

*Speech* difficulties occur in several types; there is a struggle to express what he would say, there is a sort of "plateau speech,—a type of non-melodious voice (Clark-Scripture)—a habit of coining words, and of suddenly changing the subject. The difficulty of expressing himself is perhaps the reason why the epileptic loves bombastic speech, which is the easiest way of self-expression.

It is very difficult to comprehend or define the epileptic psyche in toto. Powerful contrasts are present. We may see the highest intellect and the deepest dementia; the greatest reserve and the greatest desire to be attentive to others, especially women; depression and excitement; inaccessibility and hypocritical devotion in contrast with intense egotism (especially in religious matters); fearsome shyness and blind rage (as affects); apathy and extreme irritability. And all this is "epilepsy."

The epileptic psyche shows a distinct and complete injury to the intellect. Perception, retention, formation and association, reflection, etc., are all retarded or rendered difficult—and to such a degree that falsification occurs, so that hallucinations and fantastic ideas result. With a further degree we obtain the "twilight state" (dissociation of Ziehen). In many epileptics, however, we find normal intellect, while complete terminal dementia is rare.

The relation of psychical epilepsy to the convulsive crises and to other psychoses requires much research. The chief division of psychoses (already given) is into acute and chronic. Are these both phases of one condition? The acute form is characterized by periodicity. This seems to set up a barrier between acute and chronic forms.

"Is there really a psychical epilepsy?" "May not the psychical mani-

festations occur in non-epileptics?" This question has not yet been decided.

**Diagnosis.**—This involves study of seizures, and determination of their visible, transitory, and permanent results. We must also study the extraparoxyssmal features, the psychic behavior and the somatic concomitants in order to facilitate the recognition of the disease epilepsy.

Convulsive seizures, even when rudimentary, are readily recognized, although epileptiform convulsions may be caused by tumors of the brain, pachymeningitis, acute poisonings, progressive paresis, etc. These are no longer regarded as pictures of epilepsy. Here we come into contact with rudimentary (unilateral), or generalized convulsions proceeding from fresh or old lesions in a circumscribed portion of the cortex. It was Féré in his work on partial epilepsy, who first insisted that such convulsions represented true epilepsy as a result of organic brain disease. The greatest importance in this phase of diagnosis is bound up in operative treatment. If convulsions are really epileptiform and nothing more, the indication for operation is much more strict.

Focal phenomena in epilepsy form a link between organic and genuine epilepsy. The focal symptoms of infantile epilepsy and the same in adults present all shades of transition. In the first we see a paretic component, while in the last this is absent. One explanation is that in the grown subject a healed focus starts up convulsions but no paresis. Focal paresis is a most striking phenomenon, and may perhaps be expressed in the adult by minimal paresis or perhaps by a slight motor component (some cases of left handedness have even been attributed to such a causation). In post-paroxyssmal exhaustion, etc., the evidence of such foci may be seen. Unilateral interparoxyssmal increase of tendon reflexes and abolition or weakening of the cutaneous reflexes are to be included under exhaustion phenomena. Such are extremely important, if the unilateralism be associated with focal affections (not necessarily in the gross organic sense). Unilateral spasms cannot always be referred to a focus. In Müller's "status hemiepilepticus idiopathicus" with contractions in the Jacksonian sense, no organic basis whatever could be found. Hence there is a chance for differential diagnosis here. A number of data may be obtained from a comparison between certain focal phenomena of genuine epilepsy and Jacksonian epilepsy which would tend toward confusion. Thus several of the auras are unilateral, seen chiefly in rudimentary and abortive attacks. These auras are motor, sensible, and sensorial. Now one of these unilateral motor auras bears a close resemblance to a Jacksonian spasm, chiefly one which represents the circumscribed form, limited to one segment of a limb or one group of muscles. In this case the spasm is clonic. A distinction can be made, *i. e.*, we can only exclude the presence of an organic lesion, after a full examination of the interval period—motor weakness, depression of tactile sensibility, disturbance of coördination, etc.—the absence of which excludes the organic element. The motor aura of genuine epilepsy lasts but a few moments and is distinct from loss of consciousness. In organic cases, the motor aura is slower in development and appears only when the clonic spasms extend to other muscles. Post-paroxyssmal

paresis in the muscles first involved in the spasms speaks for organic epilepsy. The motor aura is practically uniform in each attack. Focal phenomena during a paroxysm do not as a rule indicate the source of the attack. The greatest difficulty in regard to the significance of focal phenomena is offered in cases of genuine epilepsy in which focal lesions of traumatic or syphilitic origin appear in the course of the disease.

As a rule there is no difficulty in the diagnosis of *petit mal* at the hands of an experienced practitioner. Sometimes confused with it are cases of vertigo arising from several causes (ear, stomach), migraine and syncope. Seen by themselves, these phenomena might impose themselves as *petit mal* but a little investigation will show the presence of something further of epileptic nature.

The fully developed type of *hysteria* (Charcot) is easily distinguished from epilepsy, and while the linking together of the manifestations of the seizures correspond, there is a marked difference in the motor component. The complete hysterical attack is very rare and its passionate attitudes, etc., represent a stage of delirium. Féré agrees with the Germans in that the clownish and passionate manifestations are extremely rare among the rank and file of hystericals; when present they may make the diagnosis of hysteria. Very different is it when these manifestations are absent or in the rudimentary state. Here it is extremely difficult to distinguish between hysteria and epilepsy. Féré's method of stopping an attack by pressure on the ovaries, as indicative of hysteria, is not infallible, as he himself admits. In doubtful cases we should study the interparoxysmal symptoms, which may at times point to hysteria, but often to the contrary—so that some special criterion is still in demand. This can no longer be found in the pupillary reaction, which is shown to be similar in individual cases for each disease. Neither is the temperature test of any differential value.

The "twilight state" in each disease is difficult to distinguish from its fellow, for parallelism is the evidence throughout. The chief point of differences lies in the fact that hysterical delirium is largely affective—"a delirium of recollection." In this, past experiences are sometimes re-enacted.

To sum up, the most distinguishing trait is as follows: Epilepsy in serial attacks (these also occur in hysteria) leads to pronounced psychical alterations in the sense of a defect of intelligence; and even in milder attacks injures the sound freshness and elasticity of the mind. Hysteria in every form and degree never causes any recognized decrease in the mental powers. In neurasthenia we may see affectively determined psychical inhibitory phenomena in considerable variety (irritation and deficiency symptoms) which closely resemble abortive attacks of epilepsy.

That convulsions may appear upon the foundation of neurasthenia, which are neither epileptic nor hysterical, has been shown by Oppenheim. The patients were heavily tainted, and after birth promptly showed evidence of psychopathic constitution, which crystallized within a short time into a psychasthenic character. The subjects exhibited ties, states of fear, phobias, imperative concepts and vasomotor disturbances. Upon this substratum arose, almost always as a result of affects (dis-

turbances of temper, overwork, alcohol, etc.), a condition difficult to distinguish from an epileptic seizure; it was accompanied by unconsciousness, escape of urine and feces, convulsions with tongue-biting and pupillary rigidity. Between the paroxysms occurred congestions, vertigo and phobias, with consciousness retained.

This tendency to convulsions vanishes under favorable hygienic conditions through general cures directed to combating neurasthenia and to avoidance of ordinary affective experiences. As in affect epilepsy and also affect spasms the causes are antagonistic to those of true epilepsy. This type (Oppenheim) may occur but once or twice in a lifetime, and intelligence and memory do not suffer. These cases are regarded by Oppenheim and Binswanger as representing a "*tertium quid*," a partition between psychasthenia and epilepsy which must always suggest the possibility of organic lesions.

There are many points of agreement between epilepsy and *migraine*. We see in each, periodicity, prodromes, aura phenomena (visual and auditory paresthesia), motor and sensory phenomena during a crisis, transitory psychotic states, etc. Migraine has been known to pass into epilepsy, the headache component being replaced by "scintillating scotoma." The transitions between the two are sometimes explained by the substitution of migraine for epilepsy; it appears also that migraine may mitigate epilepsy. Möbius has isolated a migraine which is merely a symptom of epilepsy, but as a rule migraine is an equivalent, not a symptom of the latter. It could well be shown that a migrainous attack might run its course in such atypical forms that to distinguish between it and an atypical form of epilepsy would be almost impossible. We may see migraine without headache or vomiting, expressing itself solely as a visual or paresthetic aura (with or without speech disturbances). Vasomotor epilepsy bears a close resemblance to migraine, since in both we may see darkening of the visual field, foggy state of consciousness, paresis; occurring in the same subject no distinction could be made.

There are two other subjects of especial interest under diagnosis: (1) To identify a man who has once been an epileptic and (2) to recognize a malingerer. In the first class the signs of a recent attack are readily recognized—such as bitten tongue, ecchymoses behind the ear or on the brow and ocular region. In case of remote attacks there should be scars on some part of the buccal mucosa, as well as on the tongue. The question of diagnosis in the interparoxysmal state is very difficult, coinciding largely with neurasthenia, which is practically the same state. Not until a seizure has occurred can a positive diagnosis be made.

In regard to *simulation*, it is easy to master the salient points and so impose on the non-expert, but the pallor of the face, dilatation of the pupil and its insensitiveness to light, the change in the pulse-rate and the cyanosis cannot be simulated. Magnan states that the simulator is unaware of the physiological spasm of the sternocleidomastoid and never simulates it. In not a few cases the simulators are themselves epileptics, which makes detection the more difficult.

**Treatment of the Epileptic Attack.**—The first and immediate measure is to determine what shall be the emergency treatment of an attack.

If it may not be obviated it is still often possible to mitigate its severity; failing in this one may still be able to prevent injury to the patient. A constant aura acts as an aid. Localized sensory or motor auras hold out the best hope, especially when, as usually is the case, they are localized in an extremity; when violent compression, flexion and extension, pulling, etc., often inhibit the attack. Forcing back the flexed thumb is in this category. But most attacks in which these phenomena occur are by nature partial.

There are certain so-called "inhibition points," pressure upon which may check an attack, such as the points of exit of the supra- and infra-orbital nerves. Certain "shock" measures are said to abort attacks, such as flogging or lashing, a blow between the shoulder blades, causing the patient to swallow a spoonful of salt, calling sharply by name. Féré, who aborted an attack by severe compression of the auricle, believed he had, so to speak, neutralized the discharge just as one paroxysmal affection may sometimes act as equivalent to another.

The practice of giving remedies to lower blood tension, to produce nausea (amyl nitrite, apomorphin) is to be condemned because they often aggravate attacks.

During a paroxysm all tight clothing is loosened, the head and chest raised, mucus removed from the mouth, and a piece of wood or rubber placed between the jaws. Nocturnal epileptics if in institutions should be watched; side pieces may be added to the bed to prevent the patient from rolling out. It is of great importance that the patient should lie undisturbed during the post-paroxysmal period. The exhaustion is made good only by rest. If awakened too soon he suffers from headache and vertigo. We should give the mind and psyche a long rest.

In serial attacks we can often arrest them in part by chloral enemata (or amylene hydrate). The same holds good for status. In cyanosis one may advise venesection. In the first stage of status cold to the head and purgatives may be given; in coma, cold affusions, or warm packs and analeptics.

**Treatment.**—1. *Hygienic-Dietetic.*—These methods are by far the most important. Such methods coincide in part with prophylaxis. The principles can be laid down in general terms only; one needs to individualize according to the age and social status of the patient. In the beginning one should seek out any sources of errors of living and whatever calls forth the frequency and intensity of the attacks, and endeavor to correct these. Special inquiry should be made, for instance, whether there be too much mental effort and stress and if such act as undue irritants. In all severe and frequent epilepsies all mental strain and school studies must be suspended at once, the criteria being such neurasthenic symptoms as headache, rapid fatigue, insomnia, and general malnutrition. In their absence one may not rightly infer that the daily mental stress or school work is not harmful, and therefore the cessation of such activities is desirable in all cases showing severe or frequent grand mal, at least during a few months of careful observation and study of the individual case. Afterward a gradual increase of educational study may be allowed for two or three hours at home. There should



be, however, long periods of rest intervals even then. Often the plan of study should be prescribed or supervised by the physician.

Not a little advantage gained may be definite information of the manner and character of mental work the patient performs. At first the schedule of study should be systematic and fairly cover the field of general education. Later the plan of study may be more specific, according to the liking and trend of interest of the individual and at the same time the scheme may be made to disclose the particular line of work or occupation the patient may follow in after life. The whole life of such individuals should be considered as a plan of comprehensive school training and treatment of his malady. If such a viewpoint is adopted many relapses after apparent arrest may be obviated. On the other hand, if epilepsy begins in adolescence (16 to 20 years) or even earlier and the disorder is not brought under subjection by treatment, many individuals may have to give up an ambitious career and conform their occupation to their disease. They should shun intellectual pursuits on general principles for even though the mind may not deteriorate, the disorder though not of a severe grade rarely permits an intellectual occupation to be pursued with great mental vigor. In those epileptic individuals who show congenital mental enfeeblement a certain industrial training should be prescribed. Those suffering even marked feeble-mindedness are benefited by the simpler sorts of industrial education. The schooling must be arranged on lines of physical labor-only.

However, the presence of frequent (daily) severe grand mal attacks at the outset or when the attacks are psychic, rudimentary or nocturnal in character, naturally reduces the question to one of active treatment with no immediate thought of education as such.

The choice of occupations is agriculture, horticulture, simple gardening, etc. These pursuits are, however, for those patients who are strong, muscular subjects. Men more highly educated and refined may take up the arts, carving, weaving, drawing, modeling, etc. Such occupations often permit the patient purely voluntary employment, to be on half time or able to come and go at will, which is the ideal work in consideration of the uncertain nature of his malady. Many of those less highly gifted and without private means cannot hope to become self-supporting and must seek institutional care and treatment where steady practical periods of varied employment are commonly supplied. Unfortunately only the larger and better endowed institutions of this sort or those under generous state supervision and maintenance supply the better class of epileptics the proper degree of classification which according to social demands and usage they crave. The herding together of the less severe epileptics with those imbecile or having serial nocturnal seizures in the public institutions cannot be too severely deprecated. If State care of epileptics is to be much further advanced careful segregation and classification of the young and unfavorable epileptics must be provided. Then, too, every means must be provided to furnish such properly arranged institutions in which recovery is the dominant idea and not merely custodial care. To this end these institutions should be furnished with a complete outfit for the treatment of nervous affections. This

is a better plan than the reception of epileptics in the great neurological institutes where diverse and discordant mental and nervous disorders are brought into harmful association. Epileptics should be isolated for their own as well as for the good of others. Unfortunately neither in this country nor abroad are there a sufficient number of well regulated private institutions for the acute cases. Economic conditions probably prevent this. The best that can be hoped for at present is to gain privacy in a well regulated family nearby some of the large cities in which the patient and his nurse may carry out such regulations in treatment and work as may be found desirable. If several such individuals are grouped in the same neighborhood the expense and labor of providing a common source of tutoring, workshops and amusements may be curtailed. Youthful epileptics may be sent to moderate altitudes (1000 to 2500 feet above sea level), which is decidedly beneficial. Travel should never be thought of as beneficial in the active treatment of epilepsy. Even when a fair degree of arrest has obtained relapses may be caused by such a course.

Bed treatment is of advantage in the severer grades of epilepsies (several attacks daily). It is not only a protection but it permits even better study and observation of the case than a larger freedom.

The *dietetic* treatment has always stood in the foreground and deservedly so, as at least three-fourths of all epileptics have some gross functional defects of the gastro-intestinal tract. As in other phases, a strict individualization of the dietetic regime of each patient is necessary. Often patients have a very poorly balanced ration, living on a single class of foodstuffs or what is more often encountered, over- or under-feeding. As a rule one has to deal with young fullblooded subjects who have a pronounced congestive habit. Thin anemic subjects are exceptional but in both classes excesses in diet cause coated tongue, foul breath, meteorism, borborygmi, constipation, diarrhœa, and many types of visceral pains. If possible studies in metabolism should precede active treatment. Whatever the previous diet may have been it should be transformed. All dietetic articles of an irritant nature such as shellfish, alcohol, tea and coffee should be eliminated. As a general rule the more or less complete exclusion of meats will be found advantageous. A purin free diet does not seem to be an asset. The habits of eating should be supervised. A fairly common diet card is as follows:

**BREAKFAST.**—*Allowed.*—Cup of cocoa, with cream and sugar. Wheat cereal with cream and little sugar. Two soft boiled eggs, or minced chicken. Rolls, toast, pulled bread, or zwieback. Maximum amount of butter, preferably unsalted. Strained honey. Orange marmalade or Dundee jam. Plenty of stewed fruit. No hot bread.

10 to 11 A.M. Choice of glass of milk (top), or milk and crackers. Cup custard, junket, egg-shake or raw eggs, chicken sandwich, malted milk, buttermilk, or lactone milk.

**LUNCHEON.**—*Allowed.*—Choice of fresh fish, chicken, lamb, mutton. Fowl of all kinds, except domestic duck or goose. Peas, beans, spinach, samp, rice, macaroni, spaghetti. Any vegetable that may be put through a puree sieve, such as turnips, carrots, etc. Boiled potatoes may be taken occasionally, and thoroughly masticated. Tender beets and oyster plant stewed. Choice of rice pudding, farina, corn starch, blanc mange, prune soufflé, tapioca, ice cream, *but no fruit ices*. Baked apple with cream, stewed figs, and prunes. Cream cheese, Camembert, pot cheese.

*Allowed Occasionally.*—Asparagus tips. Salad with French dressing.

*Not Allowed.*—Soups; liquids restricted to less than one glass. Steak, roast beef, pork, salt fish, or shell fish. Sweetbreads or kidneys. Radishes, raw celery, anchovy. Oranges, grape fruit, raw apples.

4 P.M. Same variety as at 11 A.M. with the additional choice of cocoa with sugar, or a farinaceous dessert, such as on luncheon list.

DINNER.—Same variety as for luncheon.

While liquids are restricted at meals, water may be taken freely between meals. No alcohol allowed.

It has been more or less definitely proved that the use of bromine as a remedy disturbs the chlorine metabolism, therefore salt may be reduced in the diet when the former is given. Numerous plans of diet have been constructed upon the foregoing idea. A sample is 1000 gms. of milk daily, 50 gms. of sweet butter, three eggs and from 300 to 400 gms. of bread with sodium chloride replaced by the same quantity of sodium bromide. The results from such a diet are often striking but it is difficult outside of special institutions to get such diets properly and consistently administered; besides, it is insipid to the average patient and is refused in the course of a few days' use. It is better so far as possible to select salt-free foods, but one should then guard carefully against bromism which under such a dietary often supervenes with remarkable celerity. It is a paradoxical behavior that often upon a salt-poor diet a patient becomes very sensitive to sodium chloride, so that a small dose, as in colonic flushing with normal salt solution, causes a seizure. Moreover, in such dietetic treatment a small addition of bromide precipitates an attack of bromism. Many confusing symptoms often supervene in the administration of salt-free diet and are far from being explained. It should, however, be tried in every case in which bromides are employed.

*Hydrotherapy.*—Morning bathing in lukewarm water for the delicate and cool baths (70° to 80° F.) for the robust patient followed by good frictional rubs are of essential benefit in treatment. If warm or lukewarm baths are employed a cool shower (60° to 80°) should always be given afterward. Alternating warm and cold showers of a variation of 15° to 20° may be employed when the circulation needs to be aroused, which is especially needed in all bromide therapy. Strong cold douches alone are not advised.

Electrotherapy has little or no place in the therapy of epilepsy. Gymnastics and massage are always valuable in increasing muscular strength and overcoming anemia when present. Packs and bed rest are often advisable in the overactive and too energetic epileptic.

The *mental* treatment is of first importance, not alone that he may be encouraged to renewed efforts to carry out the daily routine (a sample of which will be given later) which, because of its very monotony is most frequently abrogated in a few weeks if not thoroughly supervised, but that the patient may be taught right principles of mental hygiene. Often enough epileptics have had no proper bringing up either because of parental neglect or in the belief that their dilapidation of character is an essential part of the disease and which, if corrected, may entail more frequent and severe seizures. Hence the ideal treatment of all youthful epileptics should begin by removing them from the home and all its surroundings. They should be placed if possible in small colonies

where some fellowship with others may be enjoyed. At home they are often shunned by their social equals and this causes great depression and more frequent attacks.

Suggestion or hypnosis and their various modifications play little rôle although the occasional improvement noted when placed under secret remedies shows plainly that some suggestion may work to advantage. Psychoanalysis, although of greatest value in psychogenic convulsions (Bratz type), has yet to be carefully employed in genuine epilepsies. When this is done we may expect more light upon the exact mental training required in the treatment of the individual epileptic.

*Drug Treatment.*—The number of remedies which have been used is incalculable. This is natural both because it is such a stubborn disorder and because a cessation of attacks frequently follows any plan of treatment, by coincidence, suggestive effects, or because the remedy may possess a certain degree of sedative power necessary in the individual case. Before taking up such a list of drugs one should consider some of the conditions under which the disease improves or eliminates itself with the use of any drug. This includes not alone the plan of hygienic-dietetic regimen but the conditions under which spontaneous recoveries occur in young epileptics. In many such the diagnosis has been at fault or the employment of slight principles of corrective living have been sufficient to bring about amelioration or cure.

One may rightly ask if there are any anti-epileptic remedies *per se* that have a special or exclusive action on the disorder independent of their general utility as nerve sedatives. Valerian was the mainstay of the profession for ages or until the introduction of belladonna. Atropin is still greatly employed but usually in combination with other sedative drugs, and especially when vasomotor crises are prominent either in conjunction with grand mal attacks or interspersed between frankly convulsive seizures. In view of the fact that the number of individual attacks is usually so speedily reduced by the exhibition of bromides, even to the entire suppression of them, the hope that the drug is a real curative agent in the disease has been and still is widely held. So great has been the reliance upon the bromides that real scientific therapy has actually been delayed by this erroneous empiricism. Until a few years ago the almost wholesale use of the drug made it possible for a public clinic to count the cases showing bromide poisoning as the rule rather than the exception. Happily the recognition that there is an acquired art in the use of bromides is removing this reproach. Further, the sudden suppression of all the convulsive manifestations of epilepsy after prolonged use of bromides or by an abrupt withdrawal of the same is often followed by a serial or status period in which attacks occur continuously or are overlapping, in which a primary mortality of 50 per cent. obtains.

It is a safe rule never to begin the steady use of bromides until all other hygienic-dietetic plans have been tried. Personally I employ bromides in less than 10 per cent. of my cases. Probably the most convincing proof of the successful use of the bromides is furnished by the statistics of institutions in which the drug is presumably used with rigor and at the same time with caution. In these institutions all modi-

fications and combinations of bromides have been tested with the idea of the economy of dose, elimination of toxicity, and especially the use of adjuvants. The ordinary limit of tolerance of the drug is not far from 60 to 80 grains daily while the average dose should not be much above 30 to 40 grains daily. If these doses are not effectual the patient is not suited to bromide therapy. Usually the attacks are suppressed in greater part by such medication, for a time at least, until too great reliance is placed upon the drug and the main hygienic-dietetic regimen is allowed to fall into loose observance and the attacks recur as severely and as frequently as before. Thus one says the "drug is outworn," when in fact the other and more important principles of treatment have been disregarded. It is best to impress the patient at first, and constantly thereafter, that the real value of the treatment is non-medical in character rather than the reverse. One may thus obtain the attention of the patient to all the matters of routine of diet and hygiene in the absence of any specific drug medication. Then the bromide may be gradually increased by 1 grain a day, beginning at 10 to 15 grains daily as the minimum of effective medication. Some of the symptoms of bromide poisoning especially to be looked for are undue fatigue, drowsiness, abolition of sexual desire and the pharyngeal reflex, coated tongue, bromide rash and fetor of the breath. The undue prominence of any or all of these symptoms should cause one to lower or withdraw the bromide. It is generally held that some slight manifestations of many or all of the evidences of bromide ingestion as above outlined are necessary to show that the system is in a state of saturation, at which point the most effective use of the bromides is obtained. Personally, I prefer never to obtain the symptoms of bromism. I always employ the bromides as but secondary and adjuvant aids in the treatment of any case when sedatives are necessary. In many instances the judicious modification and use of baths, mouth cleanliness, colonic flushings, massage and physical gymnastics and dietetic principles (eggs, fruits, milk and vegetable foods) may cause the rapid disappearance of the first symptoms of bromism without altering the dose of the drug. In many severe cases of epilepsy in which exceptionally very high dosage of bromides may be desired it is possible by extra attention to the details of treatment under strict medical and nursing supervision to give patients as high as 180 or 200 grains daily without marked and untoward physical depression.<sup>1</sup> The whole plan of treatment including the bromides should be continued for years even though attacks should cease. Some authorities regard the plan of successful routine a necessary plan of life henceforth; its long continuance after attacks have ceased (for several years at least) seems to upbuild and make for a permanency of the restored normal psyche. In the absence or partial abrogation of the continued treatment one most frequently may successfully seek for the cause of relapses.

Epileptic children having a better tolerance than the adult to sedatives the dose may be held to 25 to 60 grains daily during the active phases of treatment. An epileptic, it must be remembered, does not have a greater

<sup>1</sup> Clark, *Am. Jour. Med. Sci.*, 1908, cxxxv, 94.

tolerance to sedatives than the normal individual, despite the common belief to the contrary. The bromides having a selective action for the cerebral cortex, one may note its sedation here first, in diminished time reaction and memory defects. The emotional change in irritability is also to be noted. One must remember, however, that the latter is usually more marked in the frankly established disorder to begin with and one must therefore not unduly accuse the drug. Often a moderate irritability may be increased to attacks of rage and even assaults of violence by sedatives,—a sort of bromide intoxication thus supervenes. Not infrequently in highly excitable individuals in whom the attacks have marked psychic expressions, the outburst of temper alone requires the withdrawal of sedatives. Now and then one not only experiences an increased number of attacks accompanied by insomnia, a tendency to hallucinate, but actual sensory illusions and a sort of manic excitement which usually disappears as the bromides develop their usual pronounced sedation. While the absence of corneal reflex is characteristic of the full action of bromide sedation its appearance in epilepsy may be late and vary much in its persistence, therefore it loses much of its guiding value on the exact amount of sedation at a given time. A wide variation in the time and degree of bromism shown in bromic acne also renders it not so trustworthy for the finer gauging of the action of the drug. The long detailed accounts of the severe forms of bromic cachexia have diminished in importance and significance in the literature of epilepsy since more watchful and observant plans of treatment have obtained. Suffice it to say in addition to the slighter symptoms heretofore detailed that there are stupor, loss of reflexes, ataxia, tremors and inability to write, anorexia, suppression of urine, severe constipation, slow and shallow respiration, and difficulty of swallowing. Death from aspiration-pneumonia or broncho-pneumonia has often supervened in such poisoned individuals. Obviously on the slightest suspicion of these grave symptoms all sedation should be discontinued. Contrary to what is often held the exhibition of bromides never causes permanent deterioration of the mental faculties.

Whenever the bromides are not effective, which as a rule is true sooner or later in the majority of cases, one may try the various substitutes including belladonna, digitalis, iron, opium, camphor, chloral, or their various combinations with bromides. The opium-bromide treatment is highly recommended and employed but has been of little permanent use in my hands. In patients with a positive Wassermann reaction it is not easy to reach the seat of the disease for there may be incurable arterial lesions. As a special resource a combination of bromides and iodides may be used with profit, preceded perhaps by an opium cure.

Organotherapy as yet offers no prospect of relief in epilepsy.

A sample of the daily routine under an attendant's care for a youthful individual suffering from an epilepsy of moderate severity is as follows:

Rise at 6 A.M. The patient to take either a glass of hot water or a cup of hot malted milk before or just after rising. Patient should then take five to ten minutes' active gymnastic exercises, with dumb-bells, medicine ball, Indian clubs, or simple setting-up (military) exercises.

6.30 A.M. Cold bath, plunge, or cold douche standing in warm water. Rub down with coarse towel and dress.

7.30 A.M. Breakfast as per dietary.

8.00 to 10.00 A.M. Two hours' physical exercise such as cutting wood, tennis, golf, horseback riding, bicycling, etc.

10.00 A.M. Light lunch as per dietary.

10.30 to 12.30 A.M. Study, reading, sedentary work, letter writing, etc.

12.30 to 1.00 P.M. Rest lying down, dressed.

1.00 to 2.00 P.M. Dinner as per dietary.

2.00 to 2.30 P.M. Rest sitting or half reclining.

2.30 to 4.30 P.M. Physical exercise either indoor or outdoor, shop, farm or garden. 4.30 P.M. Luncheon as per dietary.

5.00 to 6.00 P.M. Colon flushing alternating with cold pack in summer and warm in winter. 6.00 to 6.30 P.M. Rest.

6.30 P.M. Supper as per dietary. 7.30 to 8.00 P.M. Rest.

8.00 to 10.00 P.M. Games, sports, etc., in which patient may do as he pleases. 10.00 P.M. Warm bath or sponge and to bed.

In an ideal environment the patient lives away from home in a small country town with his companion-attendant in a small family. He is permitted to smoke in moderation if the habit is already contracted; no alcohol is allowed. A short attendance at religious services may be allowed once a week. Fishing, hunting and boating may be allowed under close supervision.

**Operative Treatment.**—Surgical measures are indicated only in so-called reflex epilepsy, partial Jacksonian and general traumatic epilepsy. The results here have not fulfilled the claims put forward. Temporary improvement with occasional remissions are seen often enough, but there is only a small fraction of permanent cures. We should, however, not discourage operation whenever any purely organic epileptoid condition appears to threaten the development of the epileptic alteration. All such operations are self-indicated without reference to epilepsy. Irritable scars should be excised and lesions of the skull, meninges and cortex remedied whenever found, on general principles.

## CHAPTER XVI.

### NEURASTHENIA. THE TRAUMATIC NEUROSES AND PSYCHOSES.

By CHARLES W. BURR, M.D.

#### NEURASTHENIA.

THE word neurasthenia is used with both an indefinite and a precise meaning. Unfortunately it is frequently used to designate very unlike conditions, sometimes as a cover for ignorance, and sometimes as a euphemism for a more serious word—insanity. It is often confused with hysteria and sometimes regarded as mere laziness. It may be so severe as to cause permanent disability or so slight as to be recovered from in a few weeks. Its symptoms are numerous, and its true primary cause is still a matter of dispute. The difficulties of its study are, therefore, manifold, and many of the apparent differences of opinion among writers arise from using the same word to designate different things. Notwithstanding the frequent use, indeed abuse, of the word, primary neurasthenia really is infrequent, and the diagnosis is becoming more rare as the methods of study improve, as our knowledge of the chemistry of disease grows, and as we examine patients with greater and greater thoroughness. Yet it is a really existing thing, and not a mere word. Strictly defined it includes only a condition of pathological weakness without discoverable lesion, showing itself by too rapid and too great fatigue, physical or mental, or both, emotional unbalance, and undue irritability (too great response to stimuli) of the nervous system. It means disability or inability more than perversion of function. The machine runs too slowly and weakly rather than wildly or pervertedly, although this also happens.

One thing always to be remembered is that there are two distinct types of the condition, one a primary disease and the other a secondary state. From the examination of the patient on any one day, and in ignorance of his past history, it is often impossible to determine which of the two we are dealing with. Time will always show, but it is important to make the correct diagnosis early and not late, and therefore it is important to learn the patient's personal and family history. It is more than probable that future discoveries in biological chemistry will decrease the number of cases diagnosed as primary and increase the number diagnosed as secondary, but there will remain a certain number prenatal in origin and not due to external causes arising after birth.

**Etiology.**—We shall first consider the cause of primary neurasthenia and then some of the diseases causative of secondary neurasthenic conditions. Primary neurasthenia is prenatal in origin and, in the



majority of cases, inherited. These two words, prenatal and inherited, are usually used by medical men as synonymous, but the former includes the latter. Hereditary influences are those carried by the germ cell and the sperm cell, from generation to generation, and are to be kept separate from the influences arising and acting after fusion of the cells. Prenatal influences include the hereditary plus those which, arising after conception, act upon the fetus from without. We know little about the effects of pathological prenatal influences, save in the case of gross disease, and nothing about the mechanics of heredity. We know but little of the late results (results appearing in childhood or later) of mild illness of the mother or of the embryo on the future mental and physical condition of the offspring. It is very possible, for example, although it has never been proved, that an acute febrile attack in a pregnant woman, not severe enough to cause abortion or anything noticeable at the child's birth, may yet so weaken it that the power of resistance to the stress of life will be greatly decreased. It is possible that alcoholic intoxication of either parent, otherwise healthy, at the time of conception, may have a permanently ill effect on the offspring which yet may not show itself till long after birth. It has been claimed that this has been clinically proved by the exclusion of all other possible causes, but we do not know, nor can we learn, whether acute alcoholism can affect human spermatozoa and ova, and to exclude all other causes is, to say the least, difficult. It has been claimed to have been experimentally proved that acute intoxication does affect the spermatozoa in certain lower animals. Chronic alcoholism may, by the widespread and serious tissue changes it produces in the parent, with consequent disturbance of function, undoubtedly affect the offspring.

The *fundamental* symptom of neurasthenia is inability to withstand the normal amount of stress without breakdown, shown by excessive fatigue and irritability. Stress is the sum of all the forces which act upon the individual organism and its constituent cells; and therefore includes disease, the wear and tear of life, everything affecting the emotions, mechanical work, and intellectual effort. In the primary form of neurasthenia this weakness is congenital; we come into the world with unequal powers of resistance to emotional, intellectual, physical, or chemical stress. We are only beginning to know something of the causes of inequality of ability to resist, and what we do know concerns almost altogether chemical immunity in mycotic disease. Of the causes of congenital muscular or mental weakness apart from the cases in which there is manifest gross disease we know nothing. It is possible that congenital abnormalities of function of one or several of the ductless glands may be at the root of much neurasthenia rather than any disease arising primarily in the nervous system itself, but today this is a theory rather than a proved fact. This is certain, that while the great majority of people are able to successfully resist the ordinary stress of life, there are a number who succumb, whatever the reason may be, under a burden which the majority carry well. The chief predisposing cause seems to be, in fact is, a bad heredity. In almost all cases of serious primary neurasthenia, if the family history

be known, there will be found "a bad strain in the blood." By this, of course, is not meant moral viciousness, but biological weakness. There is present some nervous instability in the family. It is a tendency which is inherited, not neurasthenia itself. Thus, neurasthenia does not necessarily beget neurasthenia, nor hysteria hysteria, but nervous or mental instability produces either itself or some allied weakness.

Further, since in nature there is always an endeavor (the limitations of language compel us to personify nature and speak of it as if it willed) to regain the normal, a very bad ancestry may beget a good posterity. Most neurasthenics, then, show something wrong in the family history. It may be general nervousness, eccentricity, or genius, at least pseudo-genius, or it may be some definite disease, as hysteria, insanity, epilepsy, gout, or alcoholism. General nervousness or mere invalidism is a more frequent ancestral trait than the various neuroses, psychoses, or organic nervous disease. This may be merely because invalidism is more frequent in general than serious organic or functional nervous disease, *i. e.*, the greater frequency may be merely arithmetical and not one of percentage. The writer is well aware that there is a tendency among certain writers and in the popular mind to minimize the effects of inheritance and to magnify those of environment, to believe that education and training can overcome all inherited evil tendencies, and that all of us could be well, strong, brilliant, and altogether normal if only we had been taught. Education can do much, and a bad environment may injure the good just as a good environment may help the bad, but an inheritance of health is still better than riches.

Some primary neurasthenics break without any discoverable exciting cause; in most, however, some unusual external stress is necessary, however slight. The average age of onset of marked symptoms is late adolescence or early maturity, but some subjects retain fair health till middle life and a few succumb in early childhood. Puberty is a critical period. Some of the boys and girls who in childhood appear bright and strong, in every way normal, at about fourteen or fifteen begin to complain of headache, backache, and mental tire, no longer do well at school, cease to take pleasure in active muscular play, and are fretful and peevish. They are really entering on a life of neurasthenia. Some die of senility while they are still mere children by the calendar. Only the healthy man's age is measured by the courses of the sun, the weakling's clock runs more quickly. The amount and nature of the stress necessary to cause breakdown vary with the individual. Some can carry a burden almost equal to the average, others reach the straining point far short of this. Mental work, even if carried to excess, provided it is not accompanied by worry, is not a frequent cause, but when worry is present, neurasthenia is frequent, and often the first symptom is inability to think consecutively and continuously. Many intellectual men are neurasthenic, but not often because of their work, and much of the world's work has been done by invalids. Much pessimism is due to what one may call neurasthenic dyspepsia, and the reader, especially if he be young and ingenuous and take the printed book too seriously, may, in turn, get a mental dyspepsia leading to physical neurasthenia. It is probable that

the reading of books of a certain type has caused more neurasthenia than the writing of them.

*Emotional stress* is a much more important factor than physical overwork. In women the countless little troubles of life and family care are very important. The common notion that only the rich and socially well placed among women have neurasthenia is absolutely false. It is more common among working women than among those who are relieved of the burden of work. This is becoming more noticeable since women have invaded the work formerly monopolized by men. School teachers and workers in stores and factories are especially susceptible. The mill demands and gets an awful tribute in the broken health of countless girls. It is not alone, not so much, the actual work that causes breakdown as the whole manner of life and the associations of the average mill girl. A few, one had almost written a great many, physicians have objected to the so-called higher education of women on the ground of injury to health, but they are regarded as old fogies and no one listens to them. Perhaps they will be listened to some time in the future. Today it is wisest not to endeavor to oppose the current of popular opinion and desire, for that is impossible, but let us not try to make Portias out of common mortals nor forget that motherhood is rather more important than science, literature, or even politics. Professional life, especially the practice of medicine, has done but little good to many of the women who have gone into it.

Child-bearing should in no way be injurious to normal women, but among those under par it is an occasional cause, as are the infectious fevers. Women are more prone to primary neurasthenia than men, but secondary, symptomatic cases seem more frequent in men. This is probably because men are still exposed to more stress of different kinds. Bad education is a great predisposing cause. The most important purpose of education, far more important than book learning, is to teach self-control, and none needs to be taught this as much as the offspring of the weak. Often, on the contrary, the neurasthenic that is to be receives the poorest education; he often learns unconsciously, from his elders in the family, to whine, to grumble, to be unwisely selfish. His ego is hardened, his altruism destroyed. He is petted and scolded and spoiled and soon made worthless. His little aches and pains are made much of, and obedience is unknown to him. The writer is not preaching brutality to children, and has no sympathy with the puritanical dogma that pleasure is sin and happiness a preparation for damnation, but one must learn to obey before he can command himself. Undue severity and lack of sympathy may do as much harm as the reverse. There are children, and often they are of the best, who are so out of tune with their human surroundings that they are driven in on themselves and live their own lives in a subjective world all of their own creation. This, however, more often leads to insanity than to neurasthenia.

*Occupations* are important causative factors. Those causing great emotional stress are most dangerous. Thus, actors, business promoters, speculators (financial, not philosophical), and clergymen of the intense type are especially liable to break down. The fault is not altogether

with the work. It is partly due to the fact that emotionalists are prone to be drawn to it. *Climate*, or rather the change from a temperate to a hot country, has an influence. Woodruff, for example, has described the neurasthenia occurring in white people living in the tropics. *Race*, too, is important. The American negro never suffered from neurasthenia till recently. But now it is of not infrequent occurrence, especially in the half-breeds. Alcohol, syphilis, and moral degeneration partly account for this, but more important is the fact that a lower race is trying to compete with a higher, and is unfit to do so. The strain is too great, and, although it seems a hard saying, the fittest to survive in the moral sense are often the unfittest in the biological sense. Apart from organic disease and bad morals, the mere struggle of the best specimens of an inferior race to attain the plane of a superior leads often to their downfall. Among Caucasians the Jews seem to be the most prone to neurasthenia. Of course in them it is not a question of racial inferiority, rather the reverse. The usual explanation given is that years of oppression have at last affected their stamina, but the cause, whatever it may be, is deeper than that. It certainly plays no part in this country, because several generations of living here does not seem to decrease the tendency. Those of us who read the Old Testament can recall some evidence that even in the days recorded there the disease was not unknown among them.

Much has been written to prove that some one thing is the cause of primary neurasthenia, indeed of all neurasthenia—floating kidney, ptosis of one or all of the abdominal organs, diseases of the genitalia in women, aberration of the sexual function, even lateral curvature and eye-strain, have each been upheld as the one and only cause of the condition. All these things are evil, and any one may be an exciting cause of nervous symptoms, and each should be looked for and if present attended to, but no one is a primary cause. That is inherent in the patient himself. The influence of trauma as a cause will be discussed elsewhere. Probably the most frequent exciting cause is the mere burden of life, the worries and frets, the great griefs and responsibilities, the dull routine, the lack of ability to get interested in the game of life, and, on the other hand, playing the game too hard. If we could be taught, and could obey the lesson, that everyone must live his life within the limitations of strength nature has put upon him, much neurasthenia would be prevented, but unfortunately few are taught till too late, and many who know are so enmeshed in the net of adverse circumstance that they cannot obey the lesson.

In *secondary* or *symptomatic* neurasthenia the cause can usually be found if sought for, but not a few serious organic diseases may, for a time at least, hide their own proper symptoms and show only weakness. The writer well remembers a man under his care when a hospital resident. The chiefs, distinguished and skilled men, diagnosed his case as neurasthenia. He complained of great general weakness and nothing more. After a rest cure he was persuaded and mildly browbeaten into taking more and more gentle exercise, longer and longer walks. He himself thought he was getting better. One day he told me he was too tired and weak to get up, and I, with the superior air of the young interne, rather

scolded him. He died before I had walked the length of the ward. The necropsy showed marked chronic myocarditis. He was not a neurasthenic, but died because his heart muscle could no longer do its work. Another patient, a man beginning to be old, lost his wife, to whom he had been devoted. He began to complain of pain in the back of his head and neck, and of general weakness. He slept poorly, ate capriciously, lost interest in all things, and began to pass large quantities of urine. He then took to his bed. Examination revealed nothing. He improved a great deal for several months, when a sarcoma of the thigh appeared, ran a very rapid course, and he died. The sarcoma had nothing to do with the early symptoms, the association of the neurasthenia and the sarcoma was purely accidental, but it was hard for the family doctor to make the patient's people believe so.

The later stages of *syphilis* may, for quite a long time, present nothing but neurasthenic symptoms, and, without a history of chancre or of previous organic symptoms, an accurate diagnosis is impossible. Given a man who presents symptoms of weakness without manifest cause, especially if his previous life has been healthy and he has borne stress well, it is important to inquire very closely into the possibility of luetic infection, and if there be any evidence whatever it is well to put him to the therapeutic test. The following is an example: A young man complained of loss of memory and inability to fix his attention, with general weakness. Examination revealed no sign of organic disease. His history, however, cleared the matter up. He had had a chancre about five years before, and later suffered from a transient apoplectic attack with aphasia, still later an epileptiform seizure with a temporary left-sided hemiplegia, and at another time an ocular palsy lasting only a short time. It is remarkable that such serious symptoms should have occurred and left no trace in the way of residual palsies, disturbance of the reflexes, etc., but such was the fact. His symptoms were not neurasthenic but due to a diffuse brain degeneration.

Sometimes, and with increasing frequency since the appearance of the present custom of surgeons in getting patients out of bed at the earliest possible moment after an operation, severe neurasthenia follows operation. The writer has not infrequently seen months lost out of the lives of patients and much suffering ensue because surgeons were stingy of a few more weeks in bed. This is true not only of women, but of men patients. Profound neurasthenia often follows typhoid fever and influenza, and in the latter especially there seems to be no relation between the severity of the primary infection, or rather of the symptoms during the fever, and that of the neurasthenia. It is notorious that two or three days of influenzal fever may be followed by weeks, even months, of convalescence, the patient feeling as miserable and being as weak as if he had passed through some long and dangerous illness.

There is a distinct type of neurasthenia, commoner among young men than young women, due to vicious sexual habits in adolescence. Not only does the habit itself do harm, but a large part of the literature about it does more harm. There is scarcely a book on the subject, and they are all sold to and read by youths, which does not do the boy reader harm

and, if he is neurotic, irreparable injury. Many a boy has been permanently injured in mind by fright after being made to believe, by reading pseudoscientific literature, that he is congenitally perverted, when really he is only the victim of a habit which can be cured. Some authors claim that sexual abstinence may be a provocative cause, but there is no foundation for such belief.

**Symptoms.**—It is very difficult to give a clear picture of the symptomatology of neurasthenia, because cases vary so in intensity and severity and because what symptoms or group of symptoms predominate depends so much on the personality of the patient. While the cardinal symptoms of rapid fatigue and undue response to stimuli are always present, the organs which show the irritability vary most markedly in different patients. This variability is so great that it has even been proposed to differentiate spinal, cerebral, gastric, and many other kinds of neurasthenia, but this is too great refinement in diagnosis, or rather it confuses the place of manifestation of symptoms with the place of origin of the disease. One organ may present the greatest number of symptoms, but the disease is general. It is impossible to give an account of the disease that will cover and include all cases. The best which can be done is to relate the symptoms and indicate the commonest combinations.

An attack is never sudden in onset, although there may be an acute outburst of severe symptoms. The physician may be told that the illness came on acutely, but investigation will always show that really the onset was slow. The great symptoms are physical and mental tire. The patient finds that she, for the disease is more frequent in women, is very easily fatigued. Molehills are mountains and the grasshopper a burden. Things that she could do before without effort now become burdensome. At first she does not think that there is much the matter. If the condition is severe she becomes unable to make any continuous muscular effort, and soon is bedridden. At the same time mental effort becomes difficult. In the beginning there is no difficulty in starting a train of thought, but in carrying it on for any length of time and in keeping the attention fixed. Finally, continuous thought becomes impossible. There are rare cases in which the weakness is entirely physical and not at all mental. There are men and women who can do continuous mental work when physically they are bedridden. But no matter how difficult it may be to think, there are never any profound perversions of thought. There are no delusions of grandeur, of self-accusation or of persecution, or, indeed, of any kind. The patient may be depressed and even fearful, being startled by the inability to think, and fearing it is the precursor of mental breakdown. Frequently there are morbid fears—a sense of impending evil, fear of closed or open places, etc.

There is usually some *emotional depression*; the patient is, as a rule, unhappy, but this is a natural though exaggerated result of the weakness. It arises occasionally from hyperconscientiousness, the patient feeling she is neglecting her duty, that she is not properly looking after her children or caring for her husband, that she ought to rise up and do her work, and that it is her own fault she does not do so. Or if she is of a different temperament she becomes peevish, fretful, fault-finding, and supremely

and childish selfish and unreasonable. When depression becomes profound and unsurmountable the patient is bordering on, if she has not reached, melancholia. A few patients maintain emotional equilibrium throughout a long attack. There is never any serious change in the moral sense, and the neurasthenic is never led by illness to acts of shameless crime, or even to slight infraction of the moral law. In severe cases which have lasted a long time there is frequently a great increase in selfishness, and this is not to be wondered at, because there are few Jobs in the world.

*Pain* or rather unusual feelings (paresthesias) are always present in the head. Occipital ache is the most usual complaint. This may come on apparently spontaneously or follow muscular effort, thought, or worry. There may also be brow ache or pain in the vertex. Some patients complain only of queer, indescribable sensations in the head. They speak of them as dragging, drawing, or straining feelings, or compare them to a tight band around the head. Slight vertigo is common. There may be hyperesthesia of the scalp, and pain may be referred to the hair. After the head the spine is the most frequent seat of pain. It may be localized or diffuse, and, as a rule, a mere touch on the skin over the backbone causes more pain than deep pressure. Although there is never anesthesia, the patient frequently suffers paresthesias in the hands and feet—pins and needles, a feeling of a tight glove, cold or hot flashes. Curiously enough, pain due to organic disease, *e. g.*, gallstone colic, may be fairly well borne. Paresthesias are often more distressing than pain. *Circulatory disturbances* are always present. The hands and feet are cold, moist, and bluish or bluish-red, or the skin is dry and scaly. There may be attacks of cardiac palpitation, a choking sensation as if the heart were in the throat, or the heart may seem to stop beating for an appreciable time. Sudden movement may cause vertigo. Although the skin is washed out and pasty looking, and the lips have lost their color, there is no true anemia; the circulation, not the blood, is at fault. There may be, and probably is, a decrease in the total quantity of blood in the body—that is a thing that cannot be measured—but there is never any great decrease in the hemoglobin percentage or in the number of red and white corpuscles. Leukocytosis is never present unless there is some complication. The explanation of the cases of apparent plethora, the red-cheeked patients, probably is a reduction in the quantity of the plasma of the blood, causing an increased blood count from concentration.

Loss of appetite may be present from the onset, come on later, or the appetite may be capricious—today ravenous and tomorrow gone. The sight of food may produce nausea, and its presence in the mouth may fail entirely to stimulate the salivary glands to action. This is one of the causes of difficulty in swallowing in neurasthenics. Food in the stomach may almost entirely fail to excite the gastric glands to secrete. After eating there may be a sense of weight in the epigastrium, a feeling of smothering and even pain. The intestines may be distended with gas. Sometimes eating increases the headache. The bowels are, as a rule, constipated, or constipation may alternate with diarrhœa. Pain may be referred to the appendix in the absence of any inflammation in it. Often there are annoying feelings in the rectum after a stool, as if the

bowel had not been emptied. The gastric atony may go on to catarrh and eating be followed by vomiting. When anorexia and indigestion are marked, emaciation always follows, and may be so great as to reduce the patient to a mere skeleton and make one fear the existence of some malignant disease or visceral tuberculosis.

Sleep is, as a rule, poor; either there is more or less insomnia or the patient dreams much and nightmare is common. The patient does not awake rested and refreshed, but weary and depressed. The morning is the worst period of the day with all neurasthenics. In mild cases there is no disturbance of menstruation, but in the more severe amenorrhœa occurs and leucorrhœa is common. There may be nervous irritability of the bladder and consequent frequency of micturition, and, on account of the habit of neurasthenics of taking too little liquid, the amount of urine voided may be below the normal and its specific gravity too high. Of course, the presence of albumin and casts indicates something other than neurasthenia. Endeavors have been made to find distinctive substances in the urine which would throw some light on the origin of the disease, but so far without success.

The deep *reflexes* are, as a rule, increased, but genuine, persistent ankle clonus can never be obtained, although quite often there is an abortive or spurious clonus shown by a few coarse and irregular vibrations of the foot when it is passively dorsally flexed. The existence of a persistent and rhythmical ankle clonus means organic disease. A tap on the knee often causes a start of the entire body. Although perhaps increased in amplitude, the knee-jerk is never spastic. It often diminishes under repeated stimulation, and when the patient is extremely weak it may be so slight as to be obtainable only on reinforcement. If the knee-jerk is absent it is well to look for other signs of organic nervous disease, for surely the absence is not caused by neurasthenia. Its absence is congenital and significant of nothing in one person in five hundred.

*Vision* and *hearing* are the two special senses most often affected. True deficiency of vision is rarely present at the beginning of an examination, but retinal tire soon comes on. The patient can read for a short time, but soon the print becomes blurred and she complains also of pain in the head and mental fatigue. Sometimes there is retinal hyperesthesia, and the patient desires to be kept in a dark room, saying light causes intense pain. Reversal of the fields of vision is a symptom of hysteria, not of neurasthenia, but rapid contraction of the fields as the examination proceeds, caused by retinal exhaustion or possibly by inability to hold attention acutely enough to appreciate slight stimuli, is not uncommon. The auditory symptoms are somewhat similar. Either the patient appears to be somewhat deaf, which is really a matter of lack of attention, or she is extremely sensitive to sounds of all kinds. There are no visual, auditory, or other hallucinations. Ringing in the ears is not very infrequent, but the subjective sound is never interpreted as bells or voices.

**Diagnosis.**—The diagnosis is largely by exclusion. It is necessary to exclude all possible gross organic disease. One of the diseases most often confused with neurasthenia and frequently associated with it is



*hysteria*. The error is of no great practical importance from a therapeutic point of view when it concerns physically weak hysterics, because the same methods of treatment are then useful in both diseases. As a matter of intellectual accuracy it is, however, well to keep them apart. Hysteria is as much abused a word as neurasthenia, and it would be well if it were used to designate only that group of symptoms including certain palsies, convulsive attacks, anesthetics, and respiratory and circulatory disturbance, with or without a certain type of purely mental symptoms. Convulsions, paralyses, and anesthetics are not symptoms of neurasthenia under any circumstances, and their presence indicates some other disease.

The early stage of chronic *insanity*, especially paresis, may look very much like simple neurasthenia. By the time a parietic comes under the care of an alienist, or is so ill that the question of sending him to a hospital is being considered, the diagnosis is usually not difficult, but in the early, the prodromal, stage it may be impossible. In the cases of paresis in which there are physical signs of organic disease from the outset, inequality of the pupils, abnormality in the iridic reflexes, scanning speech, spastic or absent knee-jerks, and intention tremor, the diagnosis is easy, because none of these occur in neurasthenia. Tremor is often seen, but it is an emotional tremor and not like that present in paresis. When the grandiose state is present the problem is easy, because no neurasthenic could be grandiose, but some parietics are depressed and even hypochondriacal. Moral degeneration, as shown by shamelessness or criminal acts, always excludes neurasthenia. The neurasthenic is often very selfish, but never morally degenerate. That is to say, the disease never causes moral degeneration. It may have existed before, and of course the neurasthenia will not cure it. In paresis there is a great lack of power of intellectual judgment even very early. In neurasthenia there is not so much lack of judgment as inability to form judgments continuously for any length of time. So far as the neurasthenic judges at all, she is prone to judge correctly. Notwithstanding all these points of differentiation, it may need several weeks' study before a correct opinion can be formed. When distinct delusions of any kind persist in a patient, he or she is insane and not merely neurasthenic. The best rule to follow is to regard every patient who shows pupillary and other reflex abnormalities as seriously ill.

Mild *hypochondria* is sometimes called neurasthenia, but when there are delusions, when the patient believes he is suffering from some non-existent disease, he is mentally diseased, and needs treatment appropriate for that and not for neurasthenia. It is sometimes very difficult to differentiate the very early stage of adolescent insanity from neurasthenia. Indeed, the onset is often so insidious that it is impossible to foretell whether the patient is going to be seriously ill or not. Always, however, there is some distinct mental perversion even at the start. The patient is a little queer, his manner changes, his behavior alters, his ideas show perversion, and there occurs some manifestly insane act. When hallucinations and delusions appear, the diagnosis is certain. Cases of mild melancholia may seem to be neurasthenia at the outset, but the emotional depression is always greater than the physical weakness. There is always,

even in the mildest cases, a sense of personal unworthiness. So soon as delusions of self-accusation are established there is no doubt the patient is insane. Certain cases of *Graves' disease* may be mistaken for neurasthenia. The classical case, with exophthalmos, goitre, and rapid heart, is easy enough to diagnose, no matter how many neurasthenic symptoms may be present, but cases occur in which the weakness seems to be before careful examination, the only symptom.

One of the difficult problems in medicine is the diagnosis of neurasthenic headache. There is a tendency among some physicians to jump to the conclusion that if a woman—with men they are more careful—complains of constant persistent headache it is not so severe as she imagines and is caused by a slight neurasthenia or “a run-down system.” This is rather dangerous, because, while head pain, or at least some strange feeling in the head, is common in neurasthenia, yet headache from other causes is much more frequent. Counting all kinds of headache, that from neurasthenia is not common. There is little in the headache itself which is characteristic except that it is apt to be described rather as a dull, queer, or strange feeling than as a sharp pain. It may be located in any part of the head, but most frequently in the occiput, the base of the brain the patient will say, and the nape. The next most frequent seat is the frontal region. When at all severe it is apt to greatly decrease ability to think and to bring on emotional depression, but these same effects follow headache from other causes. The eyes should always be examined, not only for refractive errors and muscular trouble, but also for optic neuritis and albuminuric retinitis. The writer has more than once seen a brain tumor in the early stage mistaken for neurasthenia. Typical migraine is easily diagnosed, but aberrant cases may require careful study. The nose should always be examined, for disease of it is a not very infrequent cause of headache, although not of paresthesia. Headache should never be regarded as neurasthenic unless all other causes are excluded and other symptoms of neurasthenia are present.

It is possible at first sight to mistake myasthenia gravis pseudoparalytica for neurasthenia, but in it the muscular weakness is at first, if not throughout the course of the disease, local. It begins with drooping of the eyelids, weakness of the muscles of mastication, and trouble in swallowing. The later picture is definite.

**Prognosis.**—The severity of neurasthenia varies greatly, and so much depends upon whether the patient can afford proper treatment and is so situated as to regulate and control the manner of her life that it is impossible to make any general statements as to prognosis. Taking all neurasthenics together, the outlook as to both life and cure is good. Many men and women are mildly neurasthenic for many years, and yet not only live, but do their duty in the world and not infrequently a great work. Much of the world's work has been done by invalids, many of them congenital neurasthenics. Throwing out the mild cases and counting only the severe ones, the patients who become bedridden and need strict rest cure, the danger to life is not great, but the likelihood of recurrence is extreme. If the patient be so situated that she can control the manner of her living, it is often possible for her to lead a fairly happy

and quite useful life by living according to rule, but should stress come to her, and sometimes even without stress, she may break again. Again, if the stress which caused the attack was very severe and the patient reacts well under treatment, the danger of recurrence is much less than if the stress was slight and the reaction slow. Thus, many busy, hard-working, and much worried men pass through a very severe attack, recovering completely and remaining well. The prognosis in secondary neurasthenia depends upon the nature of the primary disease. Cases following the acute infectious fevers, even though symptomatically severe, are usually entirely and permanently recovered from.

**Treatment.**—The means used to combat neurasthenia must vary with the severity of the attack. Some cases are so slight as to require nothing more than a change of scene, outdoor play, and, if possible, removal of the exciting cause, while others are so severe that they test the skill of the wisest physicians, who sometimes fail utterly in bringing about a cure. For patients seriously ill the Weir Mitchell rest treatment is the best, indeed, the only, method, and it will be described in detail. Its important elements are absolute rest in bed, food, massage, isolation, baths, and electricity. The patient is best treated away from her own home, in order to get rid of all association with the worries of housekeeping and family life. A well-regulated rest house, where there are but few other patients and where all the servants and nurses are well trained, is the ideal place, but it is also the most expensive. A not too large special hospital is the next best place, and home, as a rule, the worst. For working people, in the narrow meaning of the term, a bed in a ward of a general hospital is infinitely better than home. A very small number of patients, those in whom, although the physical weakness is great, emotional control is good, can be successfully treated at home. A well-trained and sensible nurse, not a chatterbox or a scandal-monger, but a woman of brains, tact, and strength, is in any event a necessity, because amateur or family nursing of a neurasthenic always spells failure. She must perform not only the official duties of a nurse, but by her presence relieve the monotony of isolation, for to leave a seriously neurasthenic person entirely alone for hours at a time is disastrous. To have ever with her a nagging nurse, who in her wisdom thinks neurasthenia a polite name for laziness, is still more disastrous. A nurse who is not loyal to the physician in charge is, to use the language of one of the newer pseudosciences, a potent source of "malignant animal magnetism." Her psychological influence is evil. Even a good nurse and admirable woman may be ill adapted for a particular patient. If the patient takes a dislike to her, even though to do so be entirely irrational, her usefulness is done. Friction between patient and nurse makes cure more difficult. Patients who need treatment for a long time should, for several reasons, have a change of nurses after a few months. The nurse, no matter how good, is prone to lose interest after a time or become "stale" or even nervous herself, for the work is hard and the strain severe. She may get too well acquainted with the patient, and sometimes, but this is extremely rare, an unwholesome friendship arises. Having obtained the nurse and got the patient to the rest house or hospital, the treatment begins. At

first rest should be absolute. The patient should not leave the bed except to go to the toilet. Very rarely the muscular weakness is so great that the bed-pan must be used. The bed should be neither too soft nor board-like in hardness. Feather beds are the worst, woven wire beds with hair mattresses the best. The patient should be put on a couch for an hour each forenoon while the bed is aired and freshly made up.

*Food* is an important element in the treatment. Such patients very soon after treatment is begun need large quantities of easily digested food, but at first it is best to use skimmed milk alone, giving the first day a few ounces, say three, every two hours from seven in the morning till nine at night. Next day the dose may be increased to four ounces, and soon the patient should be given two or two and a half quarts of milk daily. Many patients do not like milk, or believe that it makes them bilious, but a tablespoonful of limewater in each glass will often overcome this, or, if need be, it may be predigested. No positive rule can be laid down as to how long milk is to be the only food. Some patients thrive on it, and can be kept upon it for several months, but the majority need some other food after two or three weeks. It is then well slowly to enlarge the diet. The seven o'clock milk may be replaced by a glass of cocoa and at 8.30 she may have a breakfast consisting of a soft-boiled or poached egg, bread and butter, fruit, and milk, and after a few days dinner, in the middle of the day, never at night, may be added. It should consist of soup, some hot meat, vegetables, bread and butter, and some simple dessert. About a week later a light evening meal may be added. By this time the daily dietary consists of three meals, the mid-day one being quite large, and a glass of cocoa in the early morning and milk in the middle of the forenoon, middle of the afternoon, and at bedtime. The kind of bread used is important. Fresh bread and hot, moist rolls should never be taken by a neurasthenic. Bread a day old, toast, the crusty part of crisp rolls, or zwiebach are the best. Water is important and as milk is withdrawn should be supplied in its place. Red meat should not be eaten more than once a day. Eggs may be eaten raw, poached, or boiled, but not fried. In fact, all fried food should be prohibited. Coffee and tea should be prohibited in the earlier period of the treatment. Later, one cup of coffee with cream may be allowed at breakfast. Alcohol in all forms is harmful to all neurasthenics, and should not be given in any dose at any time.

The next important element in treatment is *massage*. If a neurasthenic is put to bed and given no physical exercise, evil is bound to result. She will become constipated and digestion, even if fairly well performed before, will be disturbed. The great good of massage is that it gives the patient exercise, and the muscles work without any fatigue on the patient's part. Of course, the work done is nothing like so great as in active exercise, but there is no need that it should be so. Massage also does good in increasing the peripheral capillary circulation and possibly in actually squeezing poisonous products out of the muscles. The best time to give it varies in different patients, but the treatment should never begin or end within an hour of a meal. Some women who sleep badly are helped to sleep by it, and with them it should be given just before

settling down for the night. For most patients about 3 p.m. is the best time. Whatever time is chosen, it should be the same hour every day, and not one day in the forenoon, the next in the afternoon, and the next at night. While the treatment is being given the patient should, during the first weeks, at any rate, remain quiet and not talk to or be talked to by the nurse. After the treatment is over she should remain quiet for one hour and not be disturbed for any reason. Some masseuses like to use cocoa oil or some other substance during massage. They are harmless, but of little if any benefit. There are a few women whom rubbing makes intensely nervous. Some have an abhorrence of being handled by anyone, and some do not dislike the treatment but get cold hands and feet and bluish lips after it. Oftentimes after a few days and by beginning with only a perfunctory rubbing abhorrence can be overcome. Rarely the treatment must be stopped altogether. At the beginning the treatment should not last more than half an hour, later on forty-five minutes to an hour. At first it should be very gentle, later more severe. Toward the middle of the course resistive movements should be added, and when the patient is up several hours a day she should begin light gymnastics. It is better to have massage given by some one other than the nurse, because she has enough to do in other ways.

*Electricity* is useful but not imperative. It, like massage, gives muscle exercise without work on the patient's part and has some good effect on the circulation. It is a mild stimulant to some patients, but this effect is largely, if not altogether, psychical. A few patients have such a nervous fear of it that it cannot be used to advantage. It should be given several hours after or before massage. A good way is to give it at eleven in the morning and massage in the afternoon. An ordinary nurse's faradic battery is all that is required. The two electrodes should be freshly covered with absorbent cotton for each treatment, wet with warm water, placed only a few inches apart over the bodies of the muscles of the arms and legs, and, the slowly interrupted current being turned on, each muscle be made to contract three or four times. The electrodes should not be placed on the tendons or the bony prominences. Care must be taken not to use a current so strong as to cause pain. The entire body should be gone over in about forty-five minutes. Sometimes the patient is made to feel much better by ending the treatment by using the rapidly interrupted current to the neck and heels. A small electrode is put on the nape of the neck and an electrode large enough to cover both feet placed on the soles. The rapid current is then turned on for ten minutes.

*Isolation* is imperative in all severe cases, but not necessary in the milder ones. Mental rest is needed quite as much as physical, and a neurasthenic is injured by the anxious faces of near relations and the useless sympathetic talk of friends who too often discuss her troubles with her. The kind friend who tells her how well she is looking and that she will recover right away if she will only make an effort is a very disturbing factor. The ills of life and petty annoyances which to her are great burdens are brought constantly before her by family talks. Sometimes more serious troubles in the home life right themselves after a time

by a judicious absence of talk and argument about them. Sometimes more wholesome marital relations, which have been lost, may be regained by the temporary separation of husband and wife. Sometimes people who care a great deal for each other react very badly on each other when one or the other is sick. For a shorter or longer time, then, it is best for the patient to see only the nurse, the doctor, and the masseuse. Notwithstanding the isolation, there is no need for the patient, as a rule, to lead a silent and entirely passive life. Soon the nurse should read to her for a little while each day. Later they may play games, and when she begins to get up she may read, write letters, see her friends, sew, embroider, make baskets, and so gradually resume her life.

At first she should have a warm sponge bath every day, preferably in the morning after the cocoa and before breakfast. Each extremity and the trunk should be bathed separately. Later, she can be given a warm and still later a cool plunge bath daily, and later she may bathe herself. In women the hair should be shampooed twice monthly. This is a little thing, but it gives comfort, and comfort hastens cure. All possible causes of so-called reflex irritation should be removed. Ocular troubles especially should be remedied. Local treatment of the genitalia, unless there be real disease present causing local symptoms, usually does more harm than good. Vaginal examination in young, unmarried women in the hope of finding some hypothetical cause for the disease is never justifiable. Such an examination is a great shock, and should never be made unless local symptoms imperatively demand it, and then it should be done under an anesthetic.

*Drugs* play little part in the rest treatment. Sometimes insomnia is so great as to require an hypnotic. Veronal is probably the best, although sometimes trional or sulphonal does better. Hyoscine in doses of  $1\frac{1}{2}$  gr. at night is sometimes useful, but, with the frequent perversity of active and powerful drugs, it occasionally excites instead of quieting the patient. A warm pack is often of service. Opium and its derivatives are always to be avoided. The old compound sumbul pill three times daily often quiets the nervous unrest. When there is much gastric atony ascending doses of nux vomica, beginning with 10 drops before meals, are useful. When intestinal indigestion and catarrh are present a pill of nitrate of silver, gr.  $\frac{1}{12}$ , immediately after meals is of great use. Constipation is often overcome by abdominal massage. When it fails the fluid extract of cascara in 15 minim (1 cc.) doses three times a day, or a single larger dose at night, is useful. Phosphate of soda before breakfast or a corresponding dose of the effervescent salt sometimes acts better. The combination of senna leaves stewed with prunes is homely but useful. Enemata are sometimes required. Many distressing abdominal symptoms are relieved by a binder.

Let me say one word more about the remedying of peripheral irritation in neurasthenia, and first as to the pelvic organs. Any serious disease should be treated in the same way as if the patient were not a neurasthenic. Surgical treatment of an infantile uterus will do no good. It is not a cause of the neurasthenia, but simply one of the signs of an ill-developed organism. Tears of the perineum and cervix should be repaired if they

cause local symptoms, but no immediate amelioration of symptoms is to be expected. Even the wildest doctrinaires have ceased to remove normal ovaries. If the ovaries are seriously diseased and in themselves need surgical treatment, they must be removed, but such removal usually has but little effect on the neurasthenia and sometimes is followed by very serious nervous symptoms lasting months or even years. This, or course, refers to operations before the menopause. After the pelvic organs the eyes are the organs most treated in neurasthenia. Any disease of them should be treated. It is undoubtedly true that eye-strain in persons prone to neurasthenia may cause many annoying and even distressing symptoms which can be remedied by proper treatment. Whether a floating kidney should be operated upon depends upon the circumstances and the symptoms in each particular case.

How long should a rest treatment last? No definite time can be set, for it depends upon the progress. There certainly should be some improvement in six weeks. The good signs are increase in weight or, if the patient has been fat always, a hardening of the fat and firmness of the muscles, and improvement in the color of the skin and mucous membranes. The pasty, oily, dry skin becomes clear, a little pink, and the oiliness passes away. The tongue becomes clean and the breath ceases to be foul. The appetite improves, and, the strength returning, the patient wants to get out of bed and do things. When the patient begins to get out of bed it should at first be only for a half-hour, then an hour, and every few days thereafter a longer time. She should gradually do more and more for herself and should be interested in something outside herself and outside the serious affairs of life. She should also begin to go out of doors, and finally take up her duties again.

There is one type of woman whom a rest cure ruins. Through very wilfulness and indolence such women want to stay indefinitely in bed, not because they are sick, they are not, but because they enjoy it. They are lazy and selfish, and love the luxury of having doctors and nurses. They enjoy the importance of being ill, and are too empty-headed to want to do anything in life except exist. They must be forced to get up—they never should have been put to bed—and slowly resume ordinary life. It is as hard, as Weir Mitchell said, to get some patients out of bed as it is to make others remain there. Each needs what the other wishes.

Recently there has arisen, or, rather, is now arising, an opinion among physicians that rest treatment is not as useful as many of us believe. The reason for this is not hard to discover. After it became popular it began to be used in many entirely unsuitable cases, and often was not carried out in proper detail. Sometimes the physician forgot that there is necessarily a psychic element in all treatment, and thought that all his duty was done when he mechanically set the machinery of treatment in order and then ceased to exercise any personal influence in the matter. Really the personality of the physician is a very important element in success. He needs to be a wise man, to know much, and to have great sympathy with the frailties of human nature. He must know when to be stern and when to be sympathetic, and, above all, he must never

lose or, at any rate, show that he has lost interest in the case. It is sometimes dangerous to show too much interest in the patient. Another reason is that in therapeutics, as in everything else, the pendulum of opinion is apt to swing first to one extreme and then to another, never reaching a place of stable equilibrium. Hence today there is coming into vogue a work cure for neurasthenia. It has a place and a large place in the treatment of nervous people. It has long been used by many men as the last stage of the rest treatment (Weir Mitchell so used it for years), and with many patients is a most excellent thing. Its fundamental idea is that much nervous disturbance depends upon the loss of interest in life and that work outside the things the patient has been in the habit of doing and along new lines will resurrect that interest and produce a saner, healthier view. All this is undoubtedly true.

The question at the outset, then, is, Does the patient need rest or work? The answer depends largely upon the physical strength or weakness of the patient and upon the character of the symptoms. When there is great neuromuscular weakness, when all physical effort causes rapid and great fatigue, when there is emaciation and serious digestive disturbance and any mental effort causes distress, the first, the imperative need is bed rest. On the other hand, when without much or any true physical weakness there is mere loss of the joy of living, mere boredom of life, with some, even great, nervous irritability and a little mental queerness, some doubt as to the affection of near relations, not very severe mental tire, and some but not severe gastric and intestinal disturbance, then work in the therapeutic sense is much more needed than rest. A total change in environment and the development of new interests are very necessary. Cases a little more severe need increased rest but not rest treatment. On the contrary, one hour of rest each afternoon is often quite sufficient. The important thing is to re-arouse interest in life, and this can best be done by inducing the patient to take up some new line of work, giving her at the same time much outdoor exercise and compelling her to take enough food at regular times. Finally, there is a large class of neurasthenic patients who are not severely ill, but yet suffer a great deal and need only modified rest treatment. Removal from home is almost always necessary for cure. No absolute rules can be laid down for the conduct of every patient. The matter must be left to the wisdom and discretion of the attending physician.

In the last few years a great deal has been written concerning the so-called *psychic* treatment of functional nervous diseases, and claims have been made that neurasthenics can be cured by such means. Indeed, the word *psychasthenia* is coming more and more in use to replace neurasthenia. Now, if neurasthenia be made to include all kinds of nervous symptoms and several types of mental aberration, if it means palsy of the will, and laziness and general nervousness, then certain cases of it can be cured by purely mental treatment. But the writer has not been discussing that sort of thing, but a serious and well-defined disease, the causes of which are deeply rooted in the patient. What part, then, can so-called psychotherapy play in its cure? Both much and little. No physician can help a neurasthenic if the patient doubts his skill or thinks



he is treating her in a merely mechanical way without human interest. He must have her confidence to get good results. She must believe he can cure her or, at any rate, help her. This is, of course, purely a psychological matter dependent upon his personality. But when we come to the technical side of psychotherapy the question is different. For example, the use of *hypnotism* as a therapeutic means is in the writer's mature and deliberate opinion productive of more harm than good, not only in neurasthenia, but in almost all diseases. It is a great shock to a woman to learn that anyone can make her unconscious and, so she believes and has learned by reading popular books and articles, subservient to another's will. If often repeated it certainly tends to weaken will power. Quite recently suggestion under hypnotism has been replaced by suggestion without hypnotism. This is really not such a mysterious process as some of the papers written about it would lead one to suppose. It really is nothing more than the acceptance on the patient's part of the truthfulness and accuracy of what the physician says and willingness to be led by him. Susceptibility to it indicates only ingenuousness and child-likeness of mind. One form of psychotherapy consists in an appeal to the religious sense of the patient. No harm and much good may and does come from this when properly and conservatively used, but unwisely and emotionally employed, and to the exclusion of proper medical treatment, it must do harm. It sometimes leads to very serious results because there are certain people who need not stimulation of the religious sense, but sedation. Dabbling in psychology, the occult, and the mysterious has injured a great many people and made not a few insane. It is wiser and safer for physician and priest each to keep within his own sphere. Somewhat allied to religious psychotherapy, but advised by men of a very different kind, is the treatment founded on the theory that neurasthenia and hysteria are based on the sexual function, and that if a patient be induced to make a confession of errors and sins in the long ago, she will be helped by the confession and speedily recover.

Neurasthenia in youths resulting from sexual abuse is not helped by the rest cure. As a rule, they are injured by it. Even though they seem to be physically very weak, it is far better to compel them to take outdoor exercise, cold baths, the simplest of food and not too much of it, than to put them to bed, stuff them, and give them time to day-dream about sexual pleasures. Often the hard life of a ranch among men who will not sympathize too much with them is the best means to a cure. They always need encouragement, and a quiet sensible talk does them a great deal of good.

A great deal of neurasthenia would be prevented if people were taught how to live properly. The robust and healthy, hard fibred, and with good ancestry, can get through this world with little care and less training, but those who come into the world handicapped need much of both. Although we suffer from the despotism of our ancestors, we can to a degree avoid their rule if we learn our own weakness and what we can do and what to avoid. So far environment may overcome heredity. The weak-fibred boy or girl can often escape neurasthenia if he or she be well environed and taught self-control and sensible habits of living; how much

to work, how much to play, what responsibilities to assume and what to decline. Further, good training may strengthen a weakling so that he can do more and better work and stand more and more stress.

### THE TRAUMATIC NEUROSES AND PSYCHOSES.

The use of steam and more recently of electricity in industrial life has created a new cause of disease, or, rather, has made an old cause, accidental injury, much more frequent. Traumatic neuroses and psychoses are not new diseases; it is probable that some of the slaves suffered from them at the building of the pyramids, but the tremendous development of machinery has increased the frequency of accidents and the horrors accompanying them. The rapid growth of the very modern belief in the responsibility of employers and the increase in the legal duties of common carriers toward their passengers and the general public have drawn great attention to these diseases and led to their close study. We shall not consider the effects of severe direct trauma to the brain, spinal cord, and nerves, but only the nervous and mental conditions resulting from accidents in which, although there may or may not have been injury to some other part of the body, the nervous system has escaped gross trauma. We shall exclude discussion of injury and fright as exciting causes of paralysis agitans, exophthalmic goitre, and Sydenham's chorea, and consider only traumatic neurasthenia, hysteria, and insanity not ascribable to serious local cerebral injury. We shall include the late results of concussion of the brain and strain of the lumbar muscles. Any discussion of treatment is unnecessary, because it is the same whether the illness be caused by trauma or not.

A great deal has been written and several theories propounded to explain the mechanism of the traumatic neuroses and psychoses. It is pretty generally accepted that they are in some way connected with disturbances in the central nervous system. Some metaphysical authors have assumed that mind exists independently of the brain, but works in association with it, the doctrine of parallelism, and attribute the nervous and mental disorders to psychic, as distinguished from cerebral, shock. Other writers hold that mind is merely a function of the brain, but that the brain is affected purely in its function without organic change, and still others believe that there are organic changes present but so minute that we cannot see them even under the microscope. But, although much has been written, we do not yet know what happens to and in the brain to produce traumatic neuroses and psychoses; we know nothing of their morbid anatomy. It may be assumed that whatever lesion does exist is in the brain. There are probably other, secondary, lesions in the abdominal viscera in neurasthenia, but they are hypothetical.

The possibility of *fraud* on the part of the person alleged to be ill makes the study of the traumatic neuroses and psychoses more difficult than that of most diseases. In ordinary practice the question of malingering is rarely important; in illness following an accident it is always to be thought of. Another difficulty is due to the fact that some writers are, unconsciously of course, biased in their opinions, because most of their

work consists in examining claimants for defendant corporations or the clients of lawyers who have claims. Sometimes the bias is not altogether unconscious, and, indeed, might be called by another name. Apart from downright fraud, there may be exaggeration of really existing symptoms, and a pessimistic physician may scare the patient to his increased hurt. Indeed, it has been claimed that there are no traumatic neuroses or psychoses, but that the conditions so named are the direct and sole result of suggestion on the part of the physician treating or examining the patient. It is not, however, the prevalent opinion, and is not likely to become such. A last difficulty is the fact that there is no complex of symptoms pathognomonic of trauma as a cause. There is no clinical picture from which one can conclude, knowing nothing of the history of the case and judging only from the physical examination and the patient's account of his symptoms at the time of the examination, that the illness is caused, and only can be caused, by trauma. In every case other causes might have produced the same effect. We can only say that the shock of an accident can produce this condition and cannot produce that.

The accidents most liable to be followed by serious results are those in which the accompanying circumstances are horrifying and those which occur suddenly, unexpectedly, and without any warning. Accidents which cause very severe physical results very often are followed by less severe results than others in which the physical injury is slight, and those occurring during sleep, and especially during drunkenness, are the least likely to be followed by serious so-called functional nervous troubles.

*Traumatic neurasthenia* is probably the most frequent of all the nervous conditions which follow accidents. It may be mild and transitory or severe and permanent. Its symptomatology differs in no way from that of neurasthenia from any other exciting cause. A frequent history is about as follows: The patient has fallen from a car which started as she got off or on, or has been knocked about in a collision, or thrown to the ground from a wagon or cart. She is dazed for a moment, or even made unconscious for a short time. There are also some bruises, or, it may be, a broken rib, a cut scalp, or a broken arm. She goes, or is taken, home and finds herself much shaken up, but with no fear of any serious results. The physical injuries heal well, but she finds that there is something wrong with her. She sleeps badly, her appetite is capricious, and digestion poor. Attacks of cardiac palpitation occur and the peripheral circulation is sluggish. She may at times faint, or fear she is going to do so. She is downhearted, cries easily, is peevish and fretful. She was a useful and sensible housewife; now she is fit for nothing. She tires very easily, and has lost interest in everything. She may blame herself for this loss of interest and struggle against it, or she may become wholly selfish and demand to be treated better by her family. She gets the feeling of being a martyr and ill used. Power of mental concentration goes. Headache comes on, especially the curious occipital ache which patients often describe as a feeling of unbearable strain rather than as pain. She becomes physically weak, and can scarcely drag herself around, and finally, it may be, is bedridden. The onset, instead of being slow, may be very acute and date from immediately after the accident. If she is a

working woman or a workingman's wife, her illness often leads to great financial difficulties in the family, and brooding over this and comparing it with former times increase her illness. If the case is of a kind that leads to a lawsuit, the expectation of appearing in court, the interviews with lawyers, the examination by experts all add to the illness. Many patients are improved or, indeed, cured soon after a verdict, no matter what it may be, not because the patients were malingerers, but because the horror of the trial is over. Some cases are made more severe by the long delays which occur before legal settlement is reached, and sometimes among the poor, in cases in which there is no doubt of the genuineness of the symptoms, proper treatment cannot be carried out because of the lack of money. Some patients present great physical weakness and rapid mental fatigue with little or no emotional unbalance.

Some cases which are commonly classified as traumatic neurasthenia ought rather to be designated by the term general nervousness. The patients suffer from general nervous unrest rather than real weakness. They often describe their condition by saying they "feel in a hurry all over." They fidget and fret and cannot get anything done because they do not know where to begin. The condition is, of course, not serious, but it is annoying and may last a long time.

*Traumatic lumbago*, which logically should not be studied here because it is caused by direct muscular strain, and hence is neither a neurosis nor psychosis, may be, because it so frequently accompanies and, indeed, aggravates neurasthenia and hysteria. It really is the result of strain of the muscles and ligaments of the back caused by violent and involuntary muscular effort made to save one's self in falling or being thrown. Force directly applied, as by a blow or by striking an object, is a less important causal factor. The pain of traumatic lumbago must not be confounded with the painful points in the spine which occur in hysteria. The lumbago involves more or less of the whole mass of lumbar muscles on one or both sides, and is shown objectively by rigidity when the patient twists upon the spine, or bends to either side or backward. In a severe case the attitude is characteristic. The spine is held stiff and straight, or bent forward from the pelvis. The lumbar muscles are in more or less rigid spasm. The patient moves slowly, not only in walking, but in using the hands or moving the head. On rising from a chair he supports the back by placing the hand on the thigh. The gait is slow and hesitating, the feet being dragged on the ground or barely raised from it. In very severe cases the pain is so great that the patient cannot walk, and this may lead to a suspicion of palsy from spinal cord disease. But there is no true palsy; the bladder and rectum are under control; there is no anesthesia; indeed, there are no signs of cord disease.

**Hysteria.**—Any one of the possible combinations of hysterical symptoms seen in non-traumatic cases may follow and be caused by trauma. No one combination proves a traumatic origin. It must never be forgotten that the presence of hysterical symptoms does not disprove the existence of organic cerebral or spinal disease. This association of hysteria and organic disease, whether it is a mere coincidence or whether, as is thought by some, a gross lesion may be an exciting cause of hysteria,

is seen apart from trauma. Thus, often in brain tumor, in women especially, in the early stages, hysterical symptoms, or symptoms taken to be hysterical, are so pronounced that the organic disease is entirely overlooked. It is especially important, for therapeutic and prognostic reasons, to carefully exclude organic disease before the diagnosis of hysteria is made. To diagnose cerebellar disease as hysterical atasia-abasia might lead to serious though different evils to patient and doctor.

The most frequent type of serious hysteria following an accident is that of palsy with anesthesia, but any type may develop. The palsy may be monoplegic, hemiplegic, or paraplegic. Sometimes the symptoms remain unchanged throughout the course of the illness, sometimes new ones are added, until finally the patient presents or has presented all or almost all the symptoms of the disease. A hysterical fit or series of fits may begin the illness or appear during its course. Sometimes the patient complains of only one symptom, but in such a case great care must be used in diagnosis, because monosymptomatic hysteria is very rare. Thus, the tremor of fright may become permanent. Tremors of this kind are sometimes followed by, or are the first symptom of, paralysis agitans. Among the rarer symptoms are the hysterical breast, bleeding breast, and true hysterical insanity. More or less neurasthenia is present in a great many hysterical patients, although many neurasthenics are not hysterical.

A common history is as follows: After an accident, in which the patient has been shocked, and sometimes quite a little time after, the patient finds trouble in walking or using the hands. There is associated with this headache, especially in the occiput, backache, points of tenderness along the spine, and a general feeling of weakness. The motor disability may go on to complete inability to use the affected parts. The palsy, instead of slowly increasing, may come on suddenly immediately or quite a little time after the accident. On examination the ordinary signs of organic palsy are not found. There is no true ankle clonus, or spasticity of the knee-jerk, but there is frequently anesthesia, it may be to slight touch only, or all sensibility may be absent. The anesthesia may involve one entire side, the palsied one if there is hemiplegia, and stop abruptly at the middle line, or both legs, or be in scattered spots of more or less regular geometrical shape, but having no relation to the distribution of the sensory nerves, or involve the hand or foot alone, having a clear-cut boundary anywhere above the wrist or ankle.

Sometimes single symptoms are complained of or are so predominant that the patient pays little attention to others which medically may be more important. For example, a woman was thrown to the floor of a car during the excitement following the blowing out of a fuse. She was taken to a hospital, where she was found to be dazed and confused, but not unconscious. She was bruised on the head and arms. After a few hours she went home by herself and went to bed. When seen by the writer, some months later, she complained only of loss of the sense of taste and smell, which she dated as arising after the accident, but could not remember how long after. Careful examination revealed no organic cause for the loss, and the general examination of the nervous system

brought to light no other signs of disease. More frequently anosmia and ageusia are merely part of a complex symptomatology.

A bleeding breast in a man who had had a penetrating wound of the chest seemed to have been caused by some slight local trouble in the nipple rather than by hysteria. When examined he complained of pain at the seat of the old wound, saying that the pain caused by bending forward prevented him doing manual work. Many months after the accident he suddenly became numb in the right arm and leg. During the examination, on milking the left breast, or rather pinching the nipple, about a quarter of a dram of blood spurted out. He had no other sign of anything more than general nervousness. A jury gave him \$5000 damages, but were said to be more influenced by the fact that he had actually sustained a penetrating wound of the chest than by his nervous complaints. Vomiting of blood, especially immediately after the accident, is much more frequent than bleeding from the breast. It may recur at intervals for some time, and is probably caused by rupture of some of the small veins of the œsophagus. Malingerers sometimes suck blood from cut gums to simulate this symptom.

**Mental Disturbances.**—Several types of *mental trouble* may follow accident. In fact, in a sense, there are always some mental symptoms associated with the nervous, because, even in neurasthenia, there is always rapid mental fatigue, an inability to do any continuous mental work, and oftentimes some, and sometimes very marked, decreased ability to form correct mental judgments, with disability or inability to decide any question. As to hysteria, it is a mental affection having physical symptoms, although it is not a true insanity. Under this heading, therefore, are considered the purely mental symptoms that may follow accident. The most frequent, probably, is mere mental *dulness*, resembling congenital stupidity. It may occur alone or be accompanied by the physical symptoms of neurasthenia. Alone it constitutes a true psychasthenia. There may be all degrees of severity. The man or woman who, before, was mentally quick, alert, and accurate in thought is now slow, dull, and inaccurate. The attention cannot be fixed for any length of time. There is marked mental inertia. The man who read much and high class literature now does not read at all or very little. The clerk who could rapidly add up a column of figures, calculate percentage quickly, almost automatically, now can scarcely do any arithmetic, not because he cannot do it if he tries hard enough, but because he cannot try hard enough for more than a few minutes. All grades can occur, from the slightest to the profound dementia in which mental power is lost entirely, for the time being, at least, and the patient must be cared for. The extreme cases are very rare, unless there has been some gross organic lesion resulting from direct and serious injury to the brain or skull, which, of course, puts them out of the class of psychoses. The milder cases are very common. Frequently it is not so much the direct result of the accident as a secondary consequence dependent upon the worry and grief resulting from inability to work. It may be unaccompanied by true physical weakness, although the mental inertia may render it impossible for the patient to make muscular effort for more than

a few minutes. There is sometimes added to the mental dulness emotional depression, which may be slight or severe.

Let us now take up the *insanities* in the more limited and restricted sense of the word. What types of insanity can and what cannot be caused by the shock of accident? Of course, the accident is merely the exciting cause; the predisposing cause is in the man, else everyone would become insane after an accident that would make anyone insane. The predisposing causes are the soil, the exciting causes the seed which grows in it. One of the most frequent questions which comes up is whether a given case of *paresis* has resulted from an accident. Now, of course, the classical cause of paresis is the combination of alcohol, syphilis, and strain and stress of life. There is no doubt that, given these as already existing, and given a concussion of the brain or a fracture of the skull, the brain injury may precipitate the occurrence of symptoms which did not exist before and would never have come into being had not the patient been subjected to some unusual stress. The question is rather different in those cases in which no gross injury to the skull or brain has occurred, but it appears that a severe shock to the nervous system can light up the disease. A patient with an already existing paresis, but in the early stage, may surely be made worse by an accident. In the later stages no harm is done because the mental powers are already too low to be influenced by shock. It is very important to determine whether the patient had the disease established before the accident. When a period of many months of good health has elapsed between the accident and the very beginning of the symptoms of paresis, and there has been no gross injury to the brain, the probabilities are that the two have no correlation.

When elderly people have sustained an accident the claim is often made that it has resulted in *senile dementia*. As a rule, elderly people do not stand sudden physical shock well, and frequently there is permanent loss of mental alertness in consequence of it. There is no doubt that occasionally it is the exciting cause of a true senile dementia. The cerebral arteries, although thickened, may be healthy enough to nourish the brain under ordinary circumstances, and the kidneys, although sclerosed, may also excrete fairly well and yet both fail, never to recover, under the stress of shock. There may be serious difficulty, however, in determining the causal relation of the accident. It is frequently not easy to discover what was the mental state of the patient before.

*Hysterical insane* states may be caused by accidents, as they may be caused by shock of any kind. The melancholic condition spoken of above may, in a susceptible person, pass into a true melancholia. States of acute confusional insanity and dementia præcox also occur. Drunkards bear shock worse than any other class of people, and may have an acutely oncoming insanity in consequence. So much for the positive side. Now as to the negative. True congenital *paranoia* is never caused by accident. It is a purely developmental disease and needs no external exciting agent for its production. It is doubtful also if true *acute mania* is ever so produced. The cases of *epileptic insanity* usually follow some direct injury to the brain, although the convulsions may not

occur for a long time after the accident, but there are usually some physical signs of the injury even at the beginning. This is not always the case, or rather at the beginning the physical signs may be slight. Thus, a man in a railroad car was struck on the side of the head with the metal end of a bell rope, which had snapped off when the car parted from the rest of the train. He was knocked down by the force of the blow and was senseless a few minutes. He recovered consciousness quickly and complained only of the pain in the bruise at the spot where he had been hit. The bruise quickly healed, but he continued to have pain in that region. Some months later he began to have general, not local, epileptiform convulsions. They were alleged by the railroad company to be hysterical, but his family physician claimed they were organic, and he was trephined at the seat of the injury and the inner table of the skull was found fractured, a spicule of bone pressing against the dura. There was a marked adhesive inflammation of the dura in the neighborhood. Whenever epileptiform, not hysterical, fits follow an accident it is always well to hunt for a local cerebral trauma.

**Diagnosis.**—The diagnosis of accident as a cause of nervous and mental disease is difficult, because in so many cases the question arises, Is the patient a malingerer? There is no short and easy way to a determination of the question. Mental alertness, knowledge, and skill on the part of the examiner, and care in investigation will always uncover fraud. Bias on the examiner's part may lead to great injustice, and this is a very important matter. The man who believes that no attention should be paid to the mere complaints of the claimant, the merely subjective symptoms, and that a diagnosis should be based solely on the objective signs, will often reach a different conclusion from him who thinks the mere statement of the claimant should be accepted at its face value. The middle path is always the safest. Undoubtedly there are many fraudulent claims, and just as undoubtedly perfectly legitimate claims are sometimes opposed in ways that are not fair. Some defendants are as tricky as some claimants.

Before a correct diagnosis can be made all the important facts in the case must be known. Especially important are the state of health of the claimant just before the accident and the nature of the accident. When a merely trifling accident has occurred, unaccompanied by any cause for shock, it is doubtful if the accident is the cause of the illness. For example, a man, about thirty-three years of age, had undoubted hysteria, which he claimed was due to striking his head against the roof of a buss which gave a sudden jolt, because of an inequality in the road. The man made no complaint at the time, continued his journey, attended to his affairs for about a month, and then had a hysterical convulsion followed by persistent hysterical symptoms. It was found that he never had had any permanent occupation, never had kept at any work in any one place for more than a few weeks, had caused his family, who were in good circumstances, much trouble by his laziness and shiftlessness, and had always excused himself on the plea of illness, making hypochondriacal complaints all the time, while, as a matter of fact, he was well nourished and strong. The trifling nature of the accident, together with his tem-



perament and character, made one disbelieve that the accident and the hysteria had anything to do with each other.

The next question is to determine the presence or absence of other causes, for example, preëxisting physical disease. Thus, a man complained that a few days after being violently thrown from a trolley car and sustaining many bruises his left knee suddenly gave way under him and he could not walk. He claimed, and no doubt honestly believed, that it was caused by the fall. As a matter of fact, he had locomotor ataxia, and the knee trouble was due to a Charcot joint. Although he had very slight ataxia of motion and station, the knee-jerks were absent, Argyll-Robertson pupil was present, and he said he had had slight pains in his legs for several years. Of course, the degeneration of the spinal cord necessary to produce all these symptoms could not have taken place in a week, no matter how severe the injury and shock of the accident might have been. The question sometimes arises in the case of men, whether their illness is not really caused by alcohol or syphilis, but both conditions usually present quite definite objective signs.

How long a time may elapse between the accident and the illness and yet the illness be the result of the accident? No time can be arbitrarily set down. Many persons who at first suffer little, later develop very serious symptoms, but the time between the onset of symptoms and the accident is never very long. This, of course, applies only to the neuroses and psychoses and not to the late effects of serious physical injury, such as concussion or fracture of the skull. In both these conditions there may be apparent recovery, and a long time after symptoms may arise as a direct consequence of a disease process in, say, the meninges, the direct result of the primary trauma. External conditions may bring on severe and sudden symptoms in these cases of old and quiescent injury. For example, a severe concussion may make it impossible for a man to do work in the heat, although in a cool place he may work very well. Traumatic epilepsy may begin years after the injury to the skull.

Certain diseases may come on suddenly at the time of the accident, and yet not be caused by it. For example, an old man fell as he was getting off a car. The evidence was that it was motionless, but nobody could tell whether he had stumbled or not. When he was picked up he was unconscious. When consciousness returned he was paralyzed on the right side and aphasic. He was old, his arteries very hard, his urine contained tube casts and a trace of albumin, and was of low specific gravity. He had no local signs of fracture of the skull, and the writer's opinion was that he most probably fell as a result of an apoplexy. In cases similar to the above, in which death occurs and an autopsy is held, if there is no fracture of the skull and there is a hemorrhage from the striate artery, it is very strong evidence that the patient fell in an apoplexy. On the other hand, the shock of an accident may, itself, produce an apoplexy.

The differential diagnosis between senility and the effects of trauma is often difficult. Shock may produce a premature senility, and, given a man who presents the signs and symptoms of senility, the diagnosis must depend upon the history of his condition preceding the accident.

If it be shown that, notwithstanding the existence of arteriosclerosis, the man was in good condition, and that dating from the accident mental and physical decay began, it is reasonable to suppose that the injury was the exciting cause.

Can an accident without severe physical injury to the head affect the *mental development* of a child? Now and again it is claimed that such an accident has caused imbecility. A nervous child is often dull and stupid and emotionally apathetic or unbalanced for some time after an accident in which there has been a large element of fright, but in the great majority of cases there is no permanent ill effect, the patient regaining emotional and mental equilibrium after a variable period. Permanent mental impairment may result in already pathologically high-strung children, but is very rare. When there has been serious concussion of the brain very severe and permanent effects may follow, but even in cases of concussion the natural resiliency of youth tends to bring about complete recovery. Slight concussion surely causes no permanent ill effects in healthy children.

Assuming that the patient is ill, and ill because of an accident, the next question is the exact nature of the illness. Is it a pure neurasthenia? If so, the patient will present more or fewer of the symptoms described of that disease. For our present purpose the most important thing is to exclude organic disease of the abdominal and thoracic viscera. This can only be done by careful and, it may be, repeated examination. Defendants sometimes claim in court that all the symptoms are subjective and that no one can tell how ill the claimant is. This is not altogether correct. A physician can tell much by the behavior, conduct, and appearance of a person as to whether he is ill or pretending.

The differential diagnosis between *hysteria* and *gross organic diseases* is important. A patient has convulsions, palsy, ataxia, or neurasthenia. Are the symptoms hysterical or organic? The classical hysterical fit is not hard to recognize. The absence of the initial cry, the unbitten tongue, the retention of control of the sphincters, with, after a fit is over, the passage of a large quantity of clear urine, the preservation of consciousness or rather its alteration, an aberration of consciousness without its abolition, the state in which the patient knows more or less well what is going on but is unable to communicate with anyone, the slow onset of the fit, permitting the patient to reach a place of safety, the absence of momentary rigidity before the clonic spasm comes on, the dramatic element, the seeming purpose in the movements, the long continuance, the absence of deep sleep after, the presence of signs of hysteria between the attacks, all point to hysteria.

The diagnosis of *hysterical palsy* may be difficult. This may be paraplegic, monoplegic, or hemiplegic. Authorities differ as to which is the more frequent. In the writer's experience brachial monoplegia, or rather hemiplegia, with much greater involvement of the arm than of the leg, has occurred most frequently, and paraplegia least. Although at first sight it may look like organic disease, the picture is never perfect. The condition of the reflexes is important. Hysteria never causes a loss of the knee-jerk. As a rule, it is much increased, but it does not

possess the qualities of the increase present in disease of the motor cortex or lateral tract. There is not the spasticity, and when the patellar tendon is tapped the whole leg is jerked, often the whole body, and the patient becomes excited. True ankle or patellar clonus is never present. There may be a few irregular and unequal to-and-fro movements of the foot when the examination is made, but true clonus, with the regularly periodical, equal, and continuous vibration of the foot, never occurs. The plantar-jerk may be absent or very marked even in an anesthetic foot; the true Babinski reflex is never present.

*Anesthesia* is an important diagnostic symptom. It is sometimes claimed to be non-existent because the patient did not know of its existence till examined, when, it is alleged, it has been brought into being by the suggestion of the physician. Now, anesthesia may be due to suggestion, but the explanation of many cases is that it is so slight as not to have made it difficult for the patient to feel objects. A patient may have tactile anesthesia sufficient to make him insensitive to a wisp of cotton or the light touch of a match stick, and yet feel perfectly when an object is pressed against or put in his hand. There are then multitudinous points of contact which may reinforce each other. A pretended analgesia can easily be discovered by sudden and unexpected pricks with a sharp instrument. This should not be done during the formal sensory examination, but before it and while the patient is off guard. Examiners sometimes forget that there may be preservation of the pain sense with tactile anesthesia. To jab a patient with a pin and because he says it hurts conclude he necessarily has no tactile anesthesia is to form a hasty judgment on insufficient evidence. The Mannkopff test, dependent upon the fact that a sudden painful stimulus frequently increases the pulse rate, is of some value in testing sensibility to pain, but sudden fright from fear of pain may do the same, and the amount of pain necessary to affect the pulse varies with the individual.

*Contractures* are frequent and may be lasting. The only muscular wasting is that from disuse. It is always diffuse, and there is never any change in the electrical reactions. When marked local wasting is present there is organic disease. It is necessary to be on guard not to mistake the general wasting of disease for trophic changes. The contractures, especially if the patient is very thin, may make the joints, notably the knee and elbow, so prominent as to create a suspicion of trophic joint trouble. Such changes do not occur, but long-continued immobility of the limb may cause a kind of adhesive inflammation around the joint and in the muscles, causing stiffness in it and shortening of the muscles. In such a case the contracture does not relax under the most profound anesthesia. The hysterical joint, which curiously is usually the knee, is swollen and painful, but shows no other sign of inflammation. There is no local heat, redness, or swollen tortuous veins. It comes on suddenly and may pass away as suddenly. Hysterical joints are sometimes claimed to be cases of tuberculous arthritis due to direct injury, and hence the importance of a differential diagnosis.

The *palsy* in hysteria may be either flaccid or rigid. In organic hemiplegia the patient in walking lifts the leg from the hip and swings

it; in hysteria the leg is dragged. Hoover has studied a sign of great value in differentiating the two. In organic hemiplegia, if the patient, lying supine, attempts to lift the palsied leg, the other is pressed down against the bed, if he lifts the non-palsied leg, the other is pressed down. This does not occur in hysteria. There is never in hysteria, a true palsy of the bladder and rectum, but temporary retention is common.

The *visual symptoms* are interesting. Contraction of the fields of vision with reversal, although common, is not pathognomonic. It occurs in brain tumor, fracture of the spine, and, indeed, not a few other diseases. A gradual contraction of the fields during the examination is characteristic rather of neurasthenia and states of exhaustion, but tubular vision apparently occurs only in hysteria. Monocular diplopia is hysterical unless due to dislocation of the lens or some other local trouble, but it must be remembered that unintelligent people often complain of monocular diplopia when really they mean they have dimmed or blurred vision in one eye. Hysterical blindness can only be diagnosed by exclusion, and must be separated from mind blindness. Since the patient avoids obstacles in walking, he may be accused of hysteria or even fraud. The condition is always accompanied by other signs of brain disease.

*Hysterical ataxia* is differentiated by the absence of the signs which accompany organic ataxia. Occasionally it is mistaken for cerebellar disease, the examiner being misled by the fact, common in the latter disease, that the patient moves the legs well in bed.

*Hysterical mutism* must be separated from aphasia and fraud. As a rule, the presence of other symptoms makes the diagnosis quite easy. The hysterical mute, as a rule, manifestly makes no real attempts to speak; the aphasic, no matter how speechless, always tries. In armies, soldiers are cured of pretended mutism by the inhalation of chloroform. When mutism is the only symptom, careful watching will soon decide.

**Prognosis.**—The danger to life is very small in any of the traumatic neuroses and psychoses. Pure neurasthenia never in and by itself leads to death, nor does it seem to increase very much the patient's susceptibility to infection if he is so situated that he can take proper care of himself. There is some dispute as to whether hysteria ever ends fatally. A few cases have been reported, but the probability of there having been an error in diagnosis is quite great. Myasthenia gravis and Landry's paralysis are sometimes mistaken for it, especially in the early stages, and occasionally patients who have had severe hysterical paralysis for years develop sclerosis of the cord and die from its secondary consequence. Whether a larger percentage develop such lesions than can be accounted for by mere arithmetical chance, or whether long-continued hysteria does predispose to sclerosis, is an unsettled question. If it does, then hysteria may indirectly be a fatal disease. Personally, if a hysterical patient should die under my care, unless from some intercurrent disease, my belief would be that there had been an error in diagnosis.

The outlook as to the duration of any given case of traumatic hysteria or neurasthenia depends on many factors. The age of the patient, the severity of the illness, the length of time it has already existed, the

ability of the patient to obtain proper treatment, are all important elements. Although some cases of hysteria are recovered from suddenly and the cure of neurasthenia is never sudden, the latter disease probably has a somewhat higher permanent recovery rate than the former, because hysterical patients are very prone to relapse when subjected to any stress or even without it. In general, old age is of bad prognostic import, because when the arteries begin to thicken and the kidneys are sclerosed the patient's power of resistance to shock decreases. Usually youth increases the chance of recovery, but hysteria may so upset the mental and nervous equilibrium as to lead to permanent disease or a life of semi-invalidism. This is, of course, especially true in those patients in whom there is a strong hereditary or congenital predisposition to hysteria. One of the most important elements in prognosis is ability of the patient to get proper treatment early in the illness, especially if it be neurasthenia. It is rarely safe to venture any prophecy as to how long illness will last. It is never safe to do so unless the physician knows all about the case. The duration of traumatic lumbago is very variable. Enough pain usually lasts after the acute stage is over to give the patient some trouble for a long time.

The prognosis of abnormal mental states is the same as when not caused by shock, except that, in general, in any mental trouble directly resulting from an external cause the patient has a better chance for recovery, other things being equal, than if it had arisen without such external cause.

## CHAPTER XVII.

### HYSTERIA.

By SMITH ELY JELLIFFE, M.D.

THE synonyms for this affection are legion. The term hysteria was in common use in the time of Hippocrates; Pliny spoke of suffocatio mulierum, Willis and Sydenham of affectio hysterica, Lorry of melancholio nervea, Sauvage and nosologists of his time of hysteric, Van Helmholt of asthma uteri, Piorry of neuropallie, while Babinski's pithiatism is the latest term proposed.

**Etiology and Psychopathology.**—No abnormal manifestation of human conduct is so well documented as that which is usually termed hysteria. From the very earliest times to the present, it has been recorded, in some of its phenomena, in all of the forms of human expression. It entered into legends and folk lore before historic records were known; it can be traced in the most ancient books of the East and West, in the poems and plays of the latter lyrical or dramatic writers, in the records of historians of events, chroniclers of action, and the makers of laws; can be seen preserved in marble in the works of the Greeks and Romans, in wood in the early Christian centuries, and in the forms of decorative art of the middle ages to the present. Priests, lawyers, philosophers, and physicians have occupied themselves with it in all ages and in all lands, and it still remains the great enigma of human personality. At no time in the past can it be said that any unanimity of opinion was reached concerning its essence; every age has known conflicting opinions, not only concerning its causes, but also its manifestations. Current notions of the times and customs of the different countries have each in their turn contributed to give it a varying presentation, but its underlying features have remained the same throughout.

Down through historical records one can trace in almost unbroken succession the development and modifications of the germ idea of an unsatisfied uterine longing as bearing a genetic relationship to hysteria, which, in the modern expressions of Freud and his disciples receives its most refined and psychologically subtle rendering as the "psychic conversion of forgotten sexual trauma." The uterine etiology died a lingering death, and had its advocates as late as the middle of the eighteenth century when the nervous system of the uterus became the chief criminal in the new pursuit.

**Humoral Doctrines.**—These doctrines were held by Galen, but a later humoralism was prevalent in the eighteenth century. Naturally there were many with mixed humoral and uterine doctrines. Auto-intoxication, gastric, intestinal, and dental, are the modern descendants.

*Nerve Theories.*—To consider the nerves as a primary seat of hysteria was a natural outgrowth, and Boerhaave sought to explain the hysterical crises by a sort of intoxication of the abdominal nerves, due to a stagnation or obstruction of the corrupting humors. Boerhaave was in reality one of the earliest suggestors of the splanchnic origin of hysteria. He did not give any better or worse definitions of these corrupting humors than many modern auto-intoxication advocates. He did not believe that the uterus was the site of the manufacture of these humors.

Although, for English-speaking students, the works of Sydenham are justly regarded as offering a remarkable picture of hysteria, yet he introduced ideas of retrogression, the influence of which is seen even at the present day. It has been noted that Galen may have believed in hysteria in the male, but, if so, such a belief was forgotten for many hundreds of years, and only about Piso's time did the presence of hysteria in the male obtrude itself upon the thoughts of clinicians. How to handle it, in view of the uterine humoral theories, was evidently difficult, and Sydenham cut the knot by saying that hypochondria was for the male what hysteria was for the female. Sydenham may also be regarded as the forerunner of the doctrine that there was no such disease as hysteria; hypochondria was everything in this century.

*Cerebral Localization Hypothesis.*—Cesbron makes the rather startling statement that from the days of Hippocrates to the nineteenth century practically only four writers located hysteria as an affection of the brain. Carolus Piso, in 1618, said that all the symptoms of hysteria came from the head and not from the uterus or intestines, and he also was one of the first to describe cutaneous anesthesia, deafness, blindness, hysterical aphonia, and hysterical tremors.

*Modern Period.*—With the development of the Charcot school the hysteria hypotheses commenced to take on more definite shape, and in the past thirty years the views show a marked similarity even though couched in different terminologies. These modern hypotheses may be grouped about three general centres, the psychological, the physiological and the biological, in all of which the psychogenic factor is prominent.

The chief *psychological* hypothesis, namely, that of dissociated personalities, received its first great impulse from Charcot himself, and has been most attractively elaborated and made popular by his pupil Janet, and even more intricately analyzed and extended by Freud and his school. For Charcot, hysteria was a psychosis, and Gilles de la Tourette is largely responsible for the prominence of the factor of suggestion in the aftercoming presentations; the extreme position of this feature alone having been advocated by Babinski, which author would seek to dismember hysteria further and give us a new grouping within this large medley, a task that Janet has also attempted, as well as Freud. Pithiatism, psychasthenia, and the anxiety neuroses are the new entities partly separated out of the hysteria conglomerate, partly from the neurasthenic mass, and partly from the initial stages of the more frank psychoses.

Of the Charcot followers, Möbius suggested the line of many later definitions. He called those morbid phenomena hysterical which were induced by ideas, and the physical as well as the psychical reactions

had a common psychogenic origin. For him every one was more or less hysterical. Every one has hysterical small coin in the bank of his personality. Gilles de la Tourette's large monograph, published in 1891, is the most faithful elucidation and amplification of the Charcot doctrine. It, with Briquet's classic, has served as the modern fount of symptomatology. At present it is recognized that the Charcot teachings were too fixed; they regarded hysteria in the light of an indivisible entity, almost in the formal light of a "species" within the limits of which were dragged a vast cohort of symptoms. Charcot described an entity—a morbus—where present-day psychiatry sees a *cohors morborum*, having really little in common, save a tendency to similar emotional reactions. The stigmata of Charcot are not alone the appanage of a definite disease, they are very widely distributed.

Probably no studies of hysteria in modern times have attracted so much attention as those of Pierre Janet, who has been so very prolific that it becomes almost impossible to give a short résumé of his standpoint. His own abstract, given at the Amsterdam Congress in 1907, is perhaps the best available. The study of somnambulism is his starting point. For him there exists in consciousness a region below, if such a term be permissible, the normal waking or personal consciousness, which is called the subconscious. Groups of ideas may exist in this, so to speak, twilight region without being at all clearly perceived by an individual, in fact, without being known at all, and yet they may operate to produce results very much as if they were the subject of voluntary attention.

The hysteric in an access of delirium lives through fancied experiences about which he knows nothing when he "comes to"—he has an amnesia for all of these events. The hysterical amnesia does not confine its manifestations to such conditions, but invades the details of life. The person who is sent on an errand forgets what she is sent for before she gets half way to her destination. This is a simple but common example. Janet would explain this by a disorder of attention. The directions are imparted to the patient but are not acutely attended to, and drop at once into the region of the subconscious and are forgotten by the waking, personal consciousness. The anesthetic arm is so because the patient does not attend sufficiently to the sensations from the arm to perceive them. There is a narrowing of the field of personal consciousness, which is but another way of expressing the defect in attention.

The synthesis of mental processes into a coherent whole constitutes the personality or ego, and the hysterical process causes a splitting, a disintegration, or a doubling of the personality. Janet's definition is that "hysteria is a form of mental depression, characterized by the retraction of the field of personal consciousness and by the tendency to the dissociation and the emancipation of systems of ideas and of functions which by their synthesis constitute the personality." For Janet the hysterical and the hypnotic states are identical, based upon the common factor of suggestibility.

The theories of Sidis, of Breur and Freud, and those of the Freud school are modified dissociation theories in which one finds more stress laid upon etiological factors. This is practically true of Freud's ideas,



which are of paramount interest to the student of mental problems since this author's work with that of Vogt's has proved a great stimulus leading to the interpretation of mental mechanisms.

Freud's hypothesis presupposes three features: (a) The rôle of psychogenesis, (b) Janet's ideas upon dissociation and psychical automatism, and (c) Binswanger's formulation of the etiological importance of the affect. As early as 1880 Breur advanced the interpretation that the individual roots of the hysterical symptoms were to be sought in ideational complexes with marked feeling tone which came about as a result of psychical or physical trauma, and in 1893 to 1895 Breur and Freud further formulated the idea that the psycho-neurotic symptoms originated from these complexes either by a process of (a) conversion, whereby the emotional (affect) excitement brought about abnormal physical innervation—this caused hysteria; or (b) by transposition of the affect through indifferent ideas (anxiety neuroses). The principle of overdetermination expressed the heaping action of an affect sufficient to determine a symptom. This same overdetermination is more concretely and physiologically expressed by Cajal's concept of avalanche action.

The ground of the conversion or the transposition lies in the immiscibility of the traumatic complex with the personality. The patient refuses to accept it, and instead of ab-reacting—and thus normally disposing of the complex—converts it or transposes it. The affect thus remains shut in or hidden in the subconscious.

Gradually Freud's attention became centred, and at first somewhat one-sidedly, upon a trauma of a genital nature. His general hypothesis is extremely intricate, and no short abstract does justice to the results obtained by his psycho-analysis, yet his present attitude may be expressed somewhat as follows: There develops, usually on a constitutional basis, in the infantile period definite sexual activities which are mostly of a perverse nature. These activities do not, as a rule, lead to a definite neurosis up to the time of puberty, which in the psychic sphere appears much earlier than in the body, but sexual fantasy maintains a perverse constellated direction by reason of the infantile sexual activities. On constitutional (affect) grounds the increased fantasy of the hysteric leads to the formation of complexes which are not taken up by the personality and by reason of shame or disgust remain buried. There therefore results a conflict between the characteristic normal libido and the sexual repressions of these buried infantile perversions. These conflicts give rise to the hysterical symptoms. It is in his contributions to the sexual theory that Freud develops his later thoughts of the sexual origin of the hysterical reaction. By sexual it is important to remember Freud is not speaking of sensual, and that sexual means the instinct of race propagation.

Inasmuch as these sexual traumata are forgotten, buried in the unconscious, it becomes necessary to dig them out by the process of psycho-analysis, either using Freud's method, or by the association tests so minutely elaborated by Jung and Riklin particularly. In practice it may take months to fully analyze some hysterical cases. When fully analyzed the patients become cured—the analysis has been a catharsis.

White<sup>1</sup> has expressed the whole matter very clearly. "The characteristic of the psychic traumata that produce hysteria is their large content of painful affect. A painful affect, fully reacted to at the time, may produce no harm, but if for any reason reaction fails, the feelings become repressed and the possibilities of dissociation are created. Failure of reaction may be due to the failure of conditions that make efficient reaction impossible, as for instance, an insult "is swallowed," a dear friend or parent is lost; and no compensation is possible. This gives rise to "retention-hysteria." Again, ideas usually of a sexual nature, which are incompatible with the personal consciousness, are repressed—ab-reaction is not permitted, no effectual catharsis takes place. This condition produces the "defense" hysteria. Finally, experiences occur in a hypnoid state—i. e., in a split-off, dissociated, or dreamy state. They produce the so-called "hypnoid-hysteria."

"The final principle of the Breur-Freud hypothesis is the principle of conversion. The strangulated affect, the unreacted-to emotion, belonging to the dissociated state which has been repressed, finds its way into bodily innervation, thus producing the motor phenomena of hysteria. In this way the strong idea is weakened by being robbed of its affect—the real object of conversion."

It is premature to pass judgment on Freud's ideas. They have their warm advocates and bitter opponents—they suggest the psychical archeologist working among the fragments of old, crumbling, and mutilated memories with a work of reconstruction in mind. The method of the archeologist who reconstructs the entire animal from a single tooth receives the approval of the archeological world, and experience is accumulating rapidly to show that the psycho-analytic method of Freud is on a sound basis in the interpretation of hysterical states.<sup>2</sup>

It has already been intimated that the superstructure built on the Charcot foundations had become a little top heavy, and the time had arrived for a more searching critique. There have been hosts of destructive critics, and the dismemberment of hysteria has been going on *pari passu* with its growth. Of the more recent serious studies looking toward a limitation of the concept and a sharpening of the diagnostic boundaries, the monograph of Steyerthal and the articles by Babinski are the most noteworthy. Steyerthal's position is extreme. For him there is no such thing as hysteria. The vast majority of the cases may be reduced to neurasthenia and mental inferiority. The stigmata of Charcot, Janet, and others are nothing but signs of tire or exhaustion. Steyerthal's exposition is ingenious and scholarly, and he has detected a number of historical errors which have been repeated for many years, but his general standpoint is too narrow. Babinski's chief position is equally restricted, but in another direction. His dismemberment of hysteria

<sup>1</sup> Current Conceptions of Hysteria, *Interstate Medical Journal*, January, 1910.

<sup>2</sup> Freud's ideas are available in Brill's very excellent translation, *Selected Papers on Hysteria and Other Psychoneuroses*, *Nervous and Mental Disease Monograph Series*, No. 4, New York, 1912. The method of Psycho-analysis by Association Tests is given best in the same writer's translation of Jung's *Psychology of Dementia Præcox*, same series, No. 3, 1909. Also in Payne's translation of Hitschmann-Freud's *Theories of the Neuroses and Psychoneuroses*, same series.

is the most complete contribution that he has given to date.<sup>1</sup> He has seen the hysterical problem grow, and with many another, he has sought to carve away from the mass the trivial and unessential and present some definite entity. This new grouping of old facts under a new head he has christened *pithiatisme*; it consists of those hysterics of tradition which respond to his formula, namely, those patients whose ills are brought into existence by the power of suggestion, and the cure of which takes place by persuasion. And suggestion, for Babinski, should express the action by which one endeavors to make another accept or realize an idea which is manifestly unreasonable, while persuasion ought to be applied to ideas that are reasonable, or which, at least, are not in opposition to good sense. All that class of phenomena which falls within the limits of this concept are truly *pithiatique*, or, using the old term which Babinski would avoid, are hysterical.

Babinski has attacked the unconscious and great extension of the idea of hysteria. The reasons for this are numerous, but may be conveniently assembled in three groups as follows: They are due to (1) errors in diagnosis; (2) to defective observation and the including in the hysterical camp of a number of simulated symptoms which do not belong there, and finally, (3) the confusion of nervous states which should be considered separately—neurasthenia, hypochondria, etc.

While all of the errors in diagnosis cannot be reviewed, Babinski calls particular attention to hemiplegia and hemianesthesia. The plantar reflex of Babinski, the combined movements of trunk and thighs, the Hoover thigh phenomenon, the sign of Grasset, the hypertonicity of paralyzed muscles, the phenomena of pronation, have come to aid in diagnosis. As for hemianesthesia, if the examination be sufficiently artful, it disappears entirely. If the more careful methods of Babinski, Stern, and Ziehen are employed it would appear that Babinski's contention is fairly true. Babinski further has pointed out that in the mistakes have been included hemoptysis, hematemesis, hematuria, and fever.

The phenomena of his second group, simulation and deceit, are difficult of classification. These, again, may be subdivided. One sub-group consists of the imitators of true *pithiatic* signs, those who simulate true hysteria—these show the various accidents such as paralyses, contractures, anesthesia, etc. This sub-group is one of the most difficult to understand, and it cannot be said that Babinski has thrown any real light on it. These have been dealt with under the heading of aggravation and exaggeration by the followers of the more classical schemes. Inasmuch as persuasion does not seem to cure them, they are, ergo, not *pithiatiques*, but simulators. Surely a very weak position. A second sub-group comprises those patients whose phenomena are fictitious and cannot be induced by suggestion. Fever and anuria are examples.

A third sub-group comprises real phenomena but consciously brought about for purposes of deception. These are the erythemata, phlyctenulæ, ecchymoses, ulcerations, sphacelæ, œdemas, etc. This group Dupré has already termed "*mythomanies*."

<sup>1</sup> *La Semaine Médicale*, January 6, 1909, No. 1, p. 1.

Finally, in discussing the third category, Babinski narrows down his conception to those cases which can be influenced by suggestion, and in a significant line he asks of the symptomatic aspect of the phenomena which suggestion can give rise to: Is it specific; does it belong to troubles related possibly to an organic affection, or to a functional malady having a mechanism different from suggestion? Here Babinski begs the whole question. For if we have a suggested chorea so like the real thing as to be inseparable by present methods, it hardly seems that his criteria help us much. Babinski in his third group separated the true pithiatiques from the "emotives"—in whom moral shock can cause phenomena to appear or disappear. He contends that in these cases an imitative factor is greater than the emotional one; finally, he erects a third sub-group in which exaggerated tendon reflexes, vasomotor phenomena, dermographia, etc., are present and usually allied with hysteria.

True hysteria then falls into three large sub-groups—the "pithiatiques," the "emotifs," and the "reflex," and these constitute a well-limited neuropathic state distinguished from all other neuroses. One can deduce that the hysterical or pithiatique phenomena depend essentially in their appearance, duration, form or disappearance, on the psychic milieu of suggestible subjects whose predisposition is susceptible of being put in play by such and such a spectacle, or such and such a proposal.

Babinski concludes that: (1) A physician should guard himself from maladroit suggestions and poor methods of examination. (2) Knowing the influence that surroundings play with these patients, these should be so modified as to get the desired effect. (3) One must avoid contrasuggestive treatment. (4) One should limit one's self in diagnosis and not make too many promises, especially in the combined cases, *i. e.*, where there are hysterical signs due in reality to organic disease. (5) From a medicolegal standpoint it is important to differentiate hysteria from simulation in order not to deprive those really suffering from just recompense on the one hand, and secondly, to limit the abuse of accident litigation in fraudulent claims.

*Physiological Theories.*—The modern physiological hypotheses of chief interest are those of Sollier, Binswanger, and the English school. They hardly do more than restate the fundamental psychological hypotheses in physiological terms. Sollier defines hysteria as a psychical functional disturbance of the brain, consisting of a torpor or a sleep of the cerebral centres, localized or generalized, temporary or permanent, and manifesting itself consequently according to the centres affected by vasomotor, trophic, visceral, sensory, motor, and final psychic disturbances, and according to its variations, its degree, and duration, by transitory crises, permanent stigmata, or by paroxysmal accidents. Confirmed hysterics are only vigilambulists whose state of sleep is more or less profound, more or less extensive. This is a dissociation hypothesis pure and simple, and inasmuch as little is known of sleep physiologically, one makes no advance by defining hysteria in terms of sleep.

Binswanger may be reckoned among the adherents of a physiological hypothesis. For him, although he claims it to be true that all hysterical phenomena can be influenced by psychical processes, yet there is no proof

that hysteria originates without exception by means of psychical processes exclusively. The disturbances of sensation and of motion are not solely of psychical origin. The same holds true for many of the visceral disturbances. The trend of recent study tends to confirm Binswanger's view.

The *hysterical change* consists in the disturbance of the orderly correlation between the psychical and material processes, and in a double direction. On the one hand for a definite series of material cortical stimuli, the parallel psychical processes are either lacking or only incompletely awakened, while, on the other hand, an excess of psychical accomplishment results in response to minimal cortical stimulation, producing a variety of back actions on the entire innervation processes which originate in the cortex, or are ruled by it. The pathological superabundance of psychical work consists not only in an increase of emotional reactions, in a sharpening of the sensations, or in the propping up of overvalued ideas, but also in the very striking facility by which the psychical processes, above all, ideas, can influence function to produce hysterical reactions.

It must be considered that in individual functional areas of the cortex, entirely different variations from the normal conditions of stimulation, both quantitatively and qualitatively, exist. At times over-stimulation, at times under-stimulation come into play, or dissimilar activities may show side by side. Furthermore, it is to be borne in mind that sharply localized disturbances of the cortex secondarily involve the capacity of other functional cortical areas, either enhancing or depressing their activities. Such functional dependencies show themselves most markedly in the disturbance of the corticomotor regions, where, for example, a "falling out" of the functional capacity of the path from cortex to bulbar or spinal motor centres causes paresis or paralysis, the incoming stimuli, influenced by anesthesia or analgesia, not being sufficiently active to cause the requisite nerve discharge leading to motor activity. This disturbance may extend to the entire nervous system, showing itself particularly in correlated physiological functional groups.

Binswanger further holds that similar dynamic disturbances are found in epilepsy, hypochondria, neurasthenia, etc., but that the distribution of the disturbed mechanism varies, and, although the entire group may be thrown into one, the psychoneuroses, yet each has its distinguishing features notwithstanding overlapping, combination, and mixed forms.

Such a physiological statement does not go far enough, however; practically the same defects may be postulated for the psychoses in general, certainly for dementia præcox at least. According to Binswanger, however, the hysterical changes are characterized by the abnormal activity of psychical influence on the innervation processes. Proportionately slight, of themselves almost negligible, cortical stimuli set up intense sensations, with pathologically increased feeling tone (pain) and conversely corticoperipheral stimuli processes, in spite of moderately strong peripheral stimuli, do not give rise to the parallel psychical processes. The intercortical or association processes show analogous disturbances. The importance of the pathological affect action has received much extended analysis and Binswanger adopts Freud's general

method of interpretation, especially with reference to the mechanism of conversion, laying less stress upon sexual traumata as the exclusive agents in the production of the reaction.

*Vasomotor Hypothesis.*—Such a view has been adopted by numbers of English neurologists, the most recent expression of which is that of Savill. For this author hysterical syncope is one of the cardinal symptoms in the disease. It represents a hysterical cerebral attack, and is due to the activity of a special reflex vasomotor centre situated in the solar plexus of the abdominal sympathetic. This is usually started into activity by an emotional stimulus from the brain, but may be started to activity and produce exactly the same kind of attack by pressure upon the groin, the stimulus passing through the iliohypogastric nerve, or by other peripheral stimulus from other areas in the skin or mucous membrane. Hysterical cerebral attacks may also be induced by variations in the vascular condition of the splanchnic area.

It is a further feature of Savill's hypothesis that the vascular conditions within the abdomen, skin, and brain oscillate among themselves, and he further drags in a modern humoralism by assuming that the symptoms are conditioned by the state of the purity of the blood. Hysterical motor disorders, paralyses, etc., are due to localized or general vascular changes in the brain; they differ from organic lesions only in extent, duration, and intensity, and in the fact that they are primarily conditioned through the sympathetic. Other sensory symptoms, hyperalgesia, etc., are due to peripheral vascular lesions, and psychogenesis may play a leading rôle.

As to the psychogenic element, Savill admitted that the mind plays a prominent part in many hysterical phenomena, but hysteria is a psychosis only in the following respects:

1. Hysterical persons throughout life present certain inherent peculiarities of mind—*e. g.*, a tendency to mental abstraction, to autohypnotism, to mental dissociation, and to dual consciousness—which render them more liable, especially on the occurrence of any disturbance of the cerebral circulation or nutrition, to exhibit abnormal mental phenomena.

2. A certain proportion of hysterical symptoms are purely mental; the mental faculties are unstable and easily disturbed, particularly the various commemorative faculties; various tricks and habits of body and mind are constantly arising.

3. The emotional side of the mind is strongly developed, and emotional outbursts which we call "hysterics" are frequent. Emotion is also a frequent determining cause of other hysterical disorders by producing vascular changes. By the well-known influence of the emotions on the vasomotor centres, vasomotor phenomena are common; and since the vasomotor centres are themselves unstable, many surprising effects and symptoms are produced as the indirect effect of emotion.

4. The mind plays an important part in *exaggerating* symptoms which have a slight physical basis, such as a vascular derangement of a part.

5. The mind also plays an important part in the *perpetuation* of symptoms after the physical basis which had initiated them has passed away.

Thus, Savill comes to a poly-etiological conclusion. No single lesion

or hypothesis is capable of explaining all the symptoms. The vasomotor system, however, plays the most important rôle, and Savill states that fully 90 per cent. of all of the symptoms are conditioned by instability of the vasomotor centres throughout the body and a want of coördination among these, associated with more or less emotional instability.

Although much is said of emotional instability, Savill makes no attempt to clear up the conception of the rôle of the emotions in this disease—a conception which practically all agree is of primary importance. If we are to succeed in analyzing hysteria we must approach the subject from the side of emotional reactivity. This is best considered here, as its final analysis will probably come from the physiological and pathological data furnished by the study of certain cellular alterations found in those psychoses with prominent disorder in the affective sphere.

A logical mode of approach is hardly possible, but in considering some of the well-known psychical reactions which accompany organic visceral disease, a breach may be made in the walls of the problem. For some it is true such phenomena are not hysterical, but if the ground is so shifted, the significance of the word is lost. These types of phenomena furnish a clue to the nature of similar phenomena.

Head has clearly presented certain mental changes which accompany visceral disease; the more patent of these may be dismissed from present consideration. In considering a special type of reflex pains of visceral disease and the influence that they consciously or unconsciously play in conduct, one approaches very closely, if not enters, the hysteria confines. Many of the phenomena called hysterical are almost constant accompaniments of organic disease of the viscera. Hallucinations of sight, of hearing, of smell, haptic hallucinations, rarely as completely developed as seen in the major psychoses, are extremely frequent. Many of these partake more of the nature of illusions, but the difference is at times an artificial one, in which connection Möbius' term of pain illusion as descriptive of the psychogenic pains of hysteria is to be considered. Moodiness is constant and leads to sleeplessness. Restlessness, attacks of crying, ideas of impending ill, feelings of exaltation, impulses to suspicion, though rarely approaching the grades usually termed delusions of reference or of observation, are extremely frequent. In all of these cases there is a type of pain which is produced by impulses passing from the affected organ up the fibres of the sympathetic, through the ganglion of the posterior root, into the central nervous system. From this result the reflex pains of the peripheral distribution of continuous segments so well described by Head. The intensity and duration of these visceral reflected pains are factors of considerable importance in the production of mental changes. Slight and evanescent pains rarely have any reaction, but the continuous, oftentimes only slight pains, are more likely to lead to the psychical effects. Women are more prone to these reflex visceral pains, and menstruation increases the severity of the mental phenomena.

Individuals thus afflicted often acquire a depressed moodiness which is, to use Head's phrase, non-projected, by which he seeks, and rather inadequately, to distinguish the state from an *emotional* state by calling

the latter projected. Consciousness is dominated by a feeling tone, but the reasoning state is not projected. The opposite side of this moodiness is an exaggerated sense of well-being. The moodiness of organic disorders may be associated, or even conditioned, by the ill-defined feelings of suspicion already mentioned. Such feelings are usually under the influence of persuasion, in which respect, again, their relation to like hysterical states is evident.

A further consideration concerns changes in attention and memory. Their analysis is extremely difficult by reason of the complexity of the phenomena. Head has reported classical examples in mild and advanced tuberculosis and the literature of organic disorders allied with hysterical reactions is replete with similar cases; some of Binswanger's unusually ample histories in his great monograph on hysteria are instances in point. Notwithstanding the difficulty of analysis, several points stand out. Fatigue is one, and the presence of the feeling due to the visceral sensations is another. Thus, attention is divided, and memory is less complete.

The individual capacities for reproducing—*i. e.*, by associated processes to bring up in consciousness memory images of feeling tone—are extremely variable, and here visuals, auditives, olfactives, affectives, etc., follow their own laws of association.

While these considerations may, at first sight, seem to have but indirect bearing upon the subject of hysteria proper, it is very evident that they do bear directly upon the mechanism by which many manifestations, usually termed hysterical, are brought into existence, and furthermore they are of importance in any attempt to sift out those cases which depend upon some definite organic factor, either from the standpoint of a visceral irritant factor in producing mental states of altered adaptation, or as causative in reducing mental resistance. The significance of Babinski's insistent critique of hysterical phenomena is amply justified by these considerations.

The next step is to ascertain how far such similar mechanisms may be set in play by lesser and lesser amounts of organic change and greater and greater amounts of mental presentation of the same class of phenomena. For it is practically coming to be conceded that the term hysteria should be restricted to the more purely psychogenetically induced disturbances of these mechanisms just considered. Not that these cover the entire series of phenomena ranged as hysterical, but they are those least capable of being understood as possibly arising as purely psychical reactions. No attempt is made to present the theories of the emotions, but it is accepted that inadequacies to emotional reactions are primary factors in the phenomena of hysteria.

Stated in very bald and primitive form, when one is brought face to face with an experience—say the oncoming of an excited animal—one's primary sensations of recognition and feeling tone of fear cause the adaptive motor mechanism of self-protection. In the healthy animal it may be to flight, but an inadequate reaction may result in inability to do anything, *i. e.*, in a paralysis, the individual components of which cannot be here analyzed. One may reason that so long as the reaction remains inadequate the individual suffers from an emotion, fear, and the



recognition of the inadequacy, *i. e.*, the paralysis, constitutes the emotion. Thus the emotion considered biologically is an evidence of maladaptation.

Head's work seems to point in the direction of the *localization* of the affected mechanisms, *i. e.*, the sympathetic, but turning to Sherrington's recent discussion of the proprio-receptive system one finds indications that the question is a much wider one and progress is being made that is eminently suggestive, at least, in its bearings on the hysteria problem.

*Biological Hypotheses.*—One terms these biological largely for the sake of convenience. They regard the hysterical reaction in the light of an adjustment to experience, or interpret it in biological terms, or perhaps in the terms of comparative psychology. The hypotheses mentioned do not conform to hard and fast classifications; thus Freud's latest interpretations could as well be classed here as under the head of psychological hypotheses. Here one can conveniently place the views of Dubois, and more particularly his pupil Schnyder, Hellpach, and Claparède, not to mention a host of others. Schnyder regards hysteria as the persistence in adult life of the childish type of reaction to the facts of life. It is a mode of reaction in persons of naïve, simple, and elementary mentality. It is a mentality lacking in development and defective in judgment and critique. Such mentalities, when placed in new environments to which they cannot adapt, or adapt with difficulty, develop the hysterical reaction. They go back to primitive methods of escaping difficulties.

Schnyder also speaks of a racial hysteria as a manifestation of the infancy of a nation, of people who are primitive and simple. It is a commonplace to note that conduct which for a white man would be called hysterical is normal for the colored race, and Kraepelin's observation of the great amount of hysteria among the natives of Java is in line with this general argument.

Modern methods of expression, the press, strikes, boycotts, unions, etc., are the safety valves of much that otherwise would be retained and repressed, and in Freud's terms "be converted" into hysterical phenomena.

Claparède lays special stress upon the fact that the hysteric shows a marked resistance to the recall of painful memories. This, viewed biologically, he considers to be a defense reaction. Suggestibility is also a protective adaptation against excessive development of personal peculiarities and tendencies, which, should they be given free play, would isolate the individual from the social world in which he lives. Instead of yielding to these impulses the suggestion of another is followed because adaptive. Suggestion thus becomes a biologically rational process—not, as Babinski would have it, a response to irrational thoughts. Claparède's ideas are interesting in view of Bleuler's very clear-cut presentation of suggestibility in the light of an adaptive mechanism with negativism as its physiological corrective, producing severally in its excess hypersuggestibility and hypernegativism in both the hysterical and dementia præcox reactions.

An attempt at a biological definition may be hazarded. Thus understood, hysteria is the carrying over into adult life of a primitive adaptive

type of nervous and mental reaction to psychical influences due to congenital anomaly, acquired development, or diminished resistance.

A few words may be said concerning the *anatomical* and *pathological* data. Here, although definite correlations have not yet been affected, there are indications which show that the factors of congenital anomaly and diminished resistance may have some demonstrable anatomical substratum. The analysis of cortical function and cellular structure is yielding results. Flechsig's data have been partly verified, corrected and amplified by the systematic study of structural variations in the cortex by cytological methods. Criteria are being established for the determination of the physiological value of cell layers and cell groups.

The layers of cells functioning for the reception of different types of stimuli are becoming fairly well delimited, but perhaps more important than all of these, pathological studies of the psychoses, dementia præcox particularly, show us, though as yet but faintly, that certain definite cell groups are affected. Here there is a most profound disturbance of the affective states with relative integrity of sensation, motion, memory, and association processes. The work of Cajal on the functional physiology of different neurones is giving more point to the ideas of dynamic polarization and some tangible anatomical details are coming to hand which give warrant for the terms "condensors" and "accumulators" of nervous energy. There is some hope of explaining the mechanism of increased discharge, lengthened discharge, and avalanche action on anatomical grounds, and problems which have always been considered as purely functional seem to offer some hope of solution by these more tangible and objective procedures.

**Symptoms.**—Before attempting a systematic presentation of the symptoms of the hysterical reaction a few remarks may be made regarding the value of this motley collection of observations in which real fragments are mingled with those born of credulity and mental laziness. The golden period of hysteria which reached such a high point as a result of the stimulus of the Charcot teachings has been followed by one of analysis in which the careful sifting of the enormous material has become an imperative necessity. One of the first striking facts of this reëxamination is the astonishing frequency of mistakes in diagnosis.

Thus, the tubercle bacillus has shown the real character of many heretofore diagnosed hysterical hemoptyses; gastric chemistry has similarly relegated many intestinal disorders into their proper position; microscopic and cryoscopic methods enable one to determine the essential features of supposed hysterical hematurias; methods of blood examination have reduced the number of hysterical fevers almost to a minimum.

In the field of the paralyses, hemiplegias, monoplegias, paraplegias, etc., mistakes have been especially frequent. With the newer signs of organic involvement of the nervous system many organic disorders are recognized which, heretofore, were called hysterical, and are even called so at the present time in their early stages. Literally thousands of sick individuals suffer from complaints which a lazy diagnosis dubs hysteria. The falsity would be revealed by a searching and intelligent analysis.

Another feature concerning the revaluation of symptoms and one of the most difficult to adequately present, is *simulation*. Unconscious simulation, if there be such, and aggravation are not included here. These are features of what must be termed the hysterical mentality. Simulation as here restricted is probably rare, yet it is constant and serves to discourage careful study of many deserving patients.

What symptoms are due to deception and deceit in the non-hysterical and in the hysterical? It is highly probable that many of those who practise such deceptions, seen from the psychiatric view point, are either debiles or mild precocious dementes. As for deliberate frauds, their symptomatology may be disregarded. There still remain a large number of symptoms which are held to be characteristic of hysteria, and which, however, are variously estimated by different observers. Here one is on the most difficult ground of the hysteria problem and each individual observer looks at the symptoms from entirely different points of view. Each reflects certain aspects of subjective truth rendering a combination impossible, hence the hopelessness of a definition.

The imperfect scales have been many. Thus, restricting the discussion purely to modern symbols, the striking motor phenomena of the "crises des nerfs," the major attacks, are the only real phenomena which should be considered as hysterical, according to the present view of Bernheim. All others are epiphenomena and are the results of education. This point of view would considerably simplify the symptomatology.

The subdivision of symptoms into two groups such as *stigmata* and *accidents* has long been current, but it is certain that in the word *stigmata* is found a relic of the methods of the middle ages in their search for trace of the presence of the Devil in accused individuals. Whereas theologians, magistrates, and medical men were in practical accord in the year 1600, at the present time there are hardly two neurologists agreed as to what shall be considered as *stigmata* or essential features of the hysterical phenomena. One knows that for Janet, anesthasias, amnesias, abulias, paralysees and changes of character represent the *stigmata*, while under the head of *accidents*, he groups hysterical attacks, somnambulism, subconscious acts, and fixed ideas, whereas Babinski claims that careful examination shows that anesthesia never exists, and between these one finds all possible variations. Some authors lay great stress on restriction of the visual fields, others practically deny its existence.

Thus for almost every symptom these conflicting attitudes of mind are found. A few uphold the absolute lack of existence of many phenomena, while others are in supreme discord as to the interpretation of phenomena which to them are subjectively true. It becomes, therefore, a difficult matter to know just how to present the symptoms. We have chosen the purely clinical method, grouping the observed symptoms according to the methods of clinical neurology and clinical psychiatry. Inasmuch as anatomy teaches the absolute interrelation of nervous structures within the entire human body, it is an idle distinction to say that hysteria is a psychosis or a neurosis. Splitting the difference by calling it a psychoneurosis simply implies that the nervous adjustments of the different parts of the body, one to another, as well as the adjustment of

the individual as a whole to his environment, show various types of disturbances called hysterical symptoms.

Hysteria is a general tendency to certain reactive expressions. The difficulty in description is an evidence of the instability of the concept, its width, and its fluctuating outlines. There lies in every person the possibility of the hysterical reaction, and it all depends either on the stimulus or the change in the resistance to bring it out. Pitt's satirical comment that "every man has his price" has its analogue in the truth that every individual has his hysterical Jack-in-the-box. Whether it will go off or not depends on the strength of the spring and the force put on the lid. When one finds a malady due to thousands of causes, it makes one conclude that either none of them has anything whatever to do with it, or that each may play a minimal rôle, and so it would seem with the hysterical personality. It may take the loss of a child to unloose the hysterical mechanism of one, while the death of a parrot is sufficient to keep some women in bed a month with an attack of hysterical paraplegia.

The aphorisms of a people reflect the aspirations of their affective lives. In the one to whom the phrase "Still waters run deep" applies, it will take a severe shock to unhinge the mechanism of affective control. So it is with the many small difficulties of life, by the one digested and forgotten, by another ever rising like the garlic of a bygone dinner and producing its disturbance in the affective sphere.

The difficulty that all students of the hysteria problem have encountered is to pick out those phenomena which are of the most significance, and which offer the most tangible lines of explanation. The various theories represent these master keys, which have seemed, for their authors at least, to have unlocked all the doors of the hysterical crypt.

**Hysterical Character.**—For didactic purposes, the symptoms of hysteria may be divided into two main classes. The foundation is a psychically abnormal character—the hysterical character, hysterical temperament—engrafted upon this are a variety of nervous and psychical phenomena due to a pathological suggestibility. Suggestibility is used here in the biological sense, not in the restricted sense as used by Babinski.

The hysterical character is chameleon-like in its variations, appearing in all classes, in the intellectual as well as the weak-minded, and baldly expressed, shows remarkable similarities to the childish type of mind. Leaving aside the minor variations, it shows itself chiefly in emotional instability or liability, in its ability to be influenced readily, in negativism and impulsiveness, in a tendency to make sensations, a remarkable egotism, desire to confabulate, to fabricate, and to simulate. Seen from the standpoint of the psychiatrist, these mental attributes stand in the foreground; the neurological manifestations are dependent upon them, and the tendency of modern analyses of hysteria is to push the psychical anomalies forward.

In the child, one finds the prototype of this character, hence some authors speak of hysterics as grown-up children, others, again, as educated savages. Students of comparative psychiatry find these features widespread in lower races. Naturally in the weak-minded one looks for much the same types, just as one expects to find them in those grown-up

individuals widely classed as degenerates or, better, as Walton suggests, deviates. The female represents in the main the physiological type of such character summaries, and Janet has made a lasting contribution to the subject in advocating the idea of a "defective synthesis," as a word picture of what occurs in those who show these traits to excess.

From this standpoint the advent of hysterical signs in comparatively healthy individuals under the influence of fatigue, of old age, disease, intoxications, of shock, and of trauma becomes comprehensible. Here, again, Janet's phrase "reduction of the mental level" is illuminating. Under such circumstances synthesis, which has been adequate to meet most of the exigencies of life, breaks down—becomes dissociated by reason of a sinking of the mental level, and permits physiologically younger and simpler types of reaction to dominate conduct. The use of the concept subconscious seems unnecessarily confusing.

Naturally the complete exhibition of these features is rarely seen in any individual case, save in the severe degenerative hysterias, and those well on the road to a hysterical psychosis. Practically one may encounter patients with well-developed hysterical symptoms, with almost complete absence of the features just enumerated, barring perhaps the element of heightened suggestibility.

**Emotional Instability.**—Excessive lability of the emotional life is most striking. The mood is constantly changing. They are readily hurt, and break into tears on the slightest reproof; again, they show a passionate scorn and relieve themselves by violent bursts of anger. Gaiety changes into depression, and prolonged periods of happiness are followed by equally moody depressions. This changeability of mood shows itself in their activities. They are happy and busy or discontented and idle, sympathetic one moment and antipathetic the next; their mood can rarely be counted on. Smiles are succeeded by frowns and friends and relatives are at a loss to explain this change, or are constantly on their guard to avert its consequences. Thus many such patients develop into "spoiled darlings." Sydenham expressed it well: "All is caprice. They love without measure those whom they will soon hate without reason."

In male hysterics particularly, one observes a greater tendency to prolonged depressions, which anomaly undoubtedly had its effect in influencing earlier writers to term these patients hypochondriacs.

**Suggestibility.**—Volumes have been written analyzing the pathological suggestibility of hystericals and science is in practical accord in acknowledging this trait as one of the most fundamental in the genesis of this mental state. It is well known that many claim it to be the master key to the whole problem, and the description of Babinski is founded upon this feature alone. True pithiatism (hysteria) for Babinski consists in those conditions which can be brought about by suggestion and cured by persuasion.

This peculiarity is to be sought in a special type of idea associations so well studied by Jung and Ricklin of recent years. In the foreground of the hysterical reaction type—for one can speak of general types only—are to be found more or less independent active idea complexes of great affect value whose development is enormously greater than in normal

individuals. The association test reactions are riddled through and through with disturbances due to these complexes, many of which are only slightly suppressed by apparently harmless reactions.

Up to the present time no adequate notion of what is meant by suggestion and suggestibility has been presented. Fortunately, psychiatry had freed itself from the notion that it is due to Zeus, Juno, Apollo, or other deity; the middle-age witchcraft and devil possession has disappeared, that is, in so far as the name is concerned. The essential ideas, however, are still compelling in the minds of the populace, but set in phrases, such as telepathy, mind influence, absent treatment, and the like. It is almost a commonplace to remark that 90 per cent. of the populace (certain cynics put it higher) do not and cannot think for themselves; they must, therefore, be led by the few, and adopt the phrases, formulas, and ideas of the few. This simply means that practically all people are more or less easily influenced or led by the suggestions of others. What is common to so many is found greatly exaggerated in a few—these are, for the most part, the hystericals, but it is only to certain types of suggested action that they are suggestible. Janet speaks of them as showing the type that leads to dissociation as a result of the narrowing of the field of consciousness.

Suggestion and suggestibility pervade the entire hysteria problem in terms of varying vagueness that are inexhaustible.

**Hysterical Negativism.**—Many studies in so-called hysterical negativism have concerned themselves with the graver psychosis, dementia præcox, but the same mechanism is met with in the hysterical. One meets it in the frequent antipathic moods, when the patients are mute, or give only monosyllabic answers, when they refuse to eat at the table, or only toy with their food, but eat large quantities of sweets in their rooms, or steal down to the cupboard late at night. One sees it in an exaggerated spirit of Christian resignation and renunciation. The extreme capriciousness and impulsiveness of the fully educated hysteric shows most exquisite negativistic phenomena.<sup>1</sup>

**Hysterical Sensationalism.**—To be constantly in the limelight, be it for purposes of attracting attention or to invite pity, is another feature. Romantic accusations, sensational confabulations, self-mutilations, and refined theatrical attempts at suicide—hysterical dream state suicidal accidents are not meant here—these have their motive power in this desire to be the observed of all observers. Lies, slandering, disfigurements, and stealing are not too much to bring this about. In the present state of journalistic enterprise in certain countries, opportunities are not wanting to gratify these ambitions. The exaggerated ego of the hysteric has always given rise to comment from the earliest times. The attention is fastened pathologically on the ego, and the constant search for sensations within the patient's own body on which to hang complaints is pursued with a definite pleasure and refined meditation. The slightest sensation is caught hold of and magnified. Somatic pains, for instance, are not created by the neurosis, but are merely utilized, aggravated,

<sup>1</sup> *Schizophrenic Negativism, Nerv. and Ment. Dis. Monograph Series.*

exaggerated, and retained by it. Careful examination, psycho-analysis, as well as physical, of so-called hysterical pains will nearly always find a nucleus of organic foundation. Freud makes the acute observation that it is always the common, the most widespread pains of humanity that seem to be most frequently called upon to play a part in hysteria.

The pains of others, however, excite but very little interest and frequently one sees jealousy and envy arise if others' pains are considered of moment in comparison. To be sick and the centre of the stage becomes a lifework, a theatrical apprenticeship, which with masterly tutelage by many physicians, usually produces a finished artist in the end. The negativistic phase, martyrdom, is a self-evident corollary.

Special attention should be directed to the tendency to confabulation. Originating in day dreaming, in the delights of fantasy, poetic fiction gradually passes through the stage of unconscious warping of truth to deliberate falsification. The will to believe becomes pathological, and a facile imagination soon supplies all of the gaps of actual observation even if it does not supply amnesia itself. Ganser's syndrome and Korsakoff's psychosis afford opportunities for studying the mechanism of hysterical confabulations of the most pronounced types.

**Motor Disturbances.**—*Attacks.*—The convulsive attacks have always attracted great attention and contributed the most striking phenomena. The capacity for this type of conduct has existed from the earliest days and it is one of the most remarkable and persistent of the features of hysteria. This transmission down through the ages, of precisely the same positions, attitudes, cries, and contortions, shows that fundamental human mechanisms are being played upon by the most primitive agents.

The striking major hysteria attacks are, however, the exception rather than the rule, and have varied so little in their manifestations that little can be added to the older descriptions. The matter of absolute frequency is a question that cannot be resolved, since there are so many factors as to make generalizations hazardous. Some physicians never see any such phenomena. It is well known how German students maintained the absence of major hysteria, while the Charcot school was richest in observations. The studies on hysteria from Germany are triple in number those of France of today. Epidemics come and go, local agitations and general excitements give rise to the proper milieu for major hysterical manifestations; the possibilities seem always there in all peoples; the environment factors may only be lacking. The religious revivals of one people are considered as manifestations of major hysteria by another and there are plenty of contemporary movements that offer excellent opportunities for studying the manifestations of major hysteria.

Statistics are notoriously unreliable in matters of this kind. Briquet, who made the first large enumeration, found that 72 per cent. of his hysterical patients (practically all women) had major hysterical attacks. Pitres, who has given a similar numerical summary, found 81 per cent. of 69 women and 22 per cent. of 31 men had major attacks. These were in the great Charcot times and under the influence of active imitative and suggestive factors. Binswanger's later figures show that 25 per cent. of 52 male patients and 54 per cent. of 80 female patients with

clearly defined hysterical symptoms suffered from convulsive attacks. Gotz in his 75 patients found 3 per cent. to be affected. Voss gives higher figures for the men, 65 per cent. of 22 men, and 57 per cent. for 101 women.

On the surface of things Slavic and Celtic races are more often affected but in any such summary due recognition should always be given to the things compared. A polyclinic population, for example, will show absolutely different statistics from a clinic or hospital population, and conditions of education, of environment, and emotional stress have more relation to these manifestations than questions of race. Behind all such statistical summaries there is a marked lack of critique of the essential determining factors, and at present there are no trustworthy factors regarding the racial relations to hysteria as seen in major hysterical attacks. The Jewish race has always shown a higher incidence than other races, but the explanation is not apparent. Personal statistical studies do not confirm the usual idea so far as the frequency of major hysterical phenomena are concerned.

During periods of intense excitement the curves of incidence of certain affections of the nervous system are bound to show marked fluctuations. Famine, war, local or general disturbance, financial distress—all of the social factors which make an impress upon the emotional nature of man, cause great perturbations in the incidence of major hysteria.

With the beginning of major hysterical phenomena in an individual the tendency to recurrence is very strong. Data are wanting as to the number of attacks, but with young individuals daily, weekly or monthly (menstrual) attacks are not uncommon. There is a natural tendency for such major manifestations to gradually become less common with advancing years. Weeks or months and finally years will go by—even in untreated individuals—with few attacks and usually only the weak-minded or trained convulsionists remain. Other manifestations may grow up, constituting the interparoxysmal or postparoxysmal modes of the same capacity for reaction.

Notwithstanding the persistence in type of the grand attacks there are countless modifications. Those accompanied by contortions may be estimated as among the severest. Many authors with the example of the epileptic convulsion in mind have sought to erect a similar march for the major hysterical attack. Such attempts are not in accord with the clinical factors, and lack of uniformity is universal. If purely general lines are sought the hysterical attacks may be described as showing three stages—a prodromal stage, one of muscular convulsions, and a poststadium.

The *prodromal stage*—one of extreme multiplicity, often, though badly, termed as *aura*—is most frequently one of mental unrest, and is shown either by restlessness, irritability, crossness, or depression; there is frequently some mental as well as motor retardation and a feeling of general distress, or a sense of tension which may lead to crying, fits of anger, or moodiness. With the fuller development a certain increase in intensity in a more restricted number of these phenomena takes place, anxiety increases, the sense of tension localizes itself more definitely, as clutchings at the throat, as palpitations, as the feeling that the skin might burst, the stomach explode, and similar feelings of tense constrict-



tion. Things dance before the patients' eyes, they hear sounds in the ears, become dizzy, and then begin to show convulsive movements. All of this may take a few days or a few minutes to develop; as a rule, the development of the earlier major attacks is longer than the later attacks, when, at times, apparently the slightest cause will precipitate an attack. In some patients the development is slow and apparently very quiet. They will lie very still for a long time—perhaps all day—in bed with the head in the pillow, and then sudden respiratory spasms, yawnings, passing of gas, etc., will indicate the presence of the severe muscular spasms. Many apparently-to-be major hysterical attacks only get to the period of the motor outbreak. They are all aura and never show the discharge phenomena.

*Convulsive Stage.*—Here the usual initial phenomena are those of extreme muscular tension, which is tonic in character. It is most frequently a slow extension movement, the head being thrown back, the arms extended, fingers extended or in a tight fist, legs straight and stiff, and toes extended. The face, rarely mask like, gives more often the impression of one in a day dream or phantasy, and the general tonus of the whole musculature is in marked contrast to that of an epileptic convulsion. The position of the body often assumes certain very characteristic attitudes, and the crucifix position is extremely common, especially among those whose associations have been extremely devout. The development of the position in point of time also shows considerable variation. Very frequently there is a progression from the head and shoulders, then to the waist, and, finally, the lower extremities, of a sort of snake-like extension that is like a gigantic overdrawn stretch. Local groups of muscles may show isolated extension positions. Some attacks consist entirely of these sinuous movements of extension with slow relaxations, and then slow extensions. The ancients, and many moderns, always interpreted these as having a strong sexual element.

In others the extension period is of but a few seconds' duration. Then the muscles relax, and a period of irregular contortional movement follows. It is not a true clonic spasm, the movements are too irregular, with rolling, tossing, kicking, and frequently screaming, with frequent recurrences of the tonic position, with opisthotonos, and then a recurrence of the rolling and individual clonic muscle movements. The tonic phases are much more coördinated than in those of relaxation—clownism is the proper application. Here a vast variety of individual muscular positions are manifest, boring of the head into the pillow, shaking it from side to side, threshing the arms backward and forward, rocking the hips, sudden sitting up and rocking backward and forward, or side to side, with hands tightly clasping the strongly flexed knees. Sometimes again the patient stands up and hops from foot to foot, or glides about, sometimes slowly and dreamily, with eyes widely opened, turned upward and arms extended, or suddenly breaks into a wild dance. In most of these cases the dramatic or theatrical character of the impersonation is very remarkable. The individual variations are too numerous to mention. Some of the antics of dementia præcox (katatonic) patients have been described as being those of major hysteria.

The facial expression often shows in these various phases a similar changeability, sometimes devoid of any expression, again it takes on the character of one in great joy or hatred, or anger; it leers and squints, pouts and disdains. It is dramatic or ecstatic, repulsive or enticing, and psycho-analysis often reveals that the idea associations passing in the mind are such as are expressed by such facial movements.

This period shows extreme variability not only in the form of muscular expression, but also in point of time. There is constant change in many patients, while in others monotony is the characteristic stamp. Some attacks come and go, others are over with in one explosion, but most attacks last some time—rarely less than fifteen to thirty minutes, more often one to two hours, frequently several hours, and in rare instances, days. Some hysterical dream states extend over months—if psychiatry is advanced enough to assure the differentiation from some abortive forms of dementia præcox or certain confused manic-depressives.

Notwithstanding the statements of many of the Charcot school that the major hysterical attack begins with a cry, it would appear that such a mode of onset is comparatively rare. It probably was an imitative phenomena in France when the great cultivation of hysteria was in progress. The hysterical cry has rarely the same sound as that of the initial cry of the epileptic attack. Accidents not infrequently happen in the stormy attacks; the tongue may be wounded and a cyanotic color of the lips is frequent. Not much dependence can be placed on the presence or absence of mucus. Absolute increase in the production of saliva is rare.

Ordinary hysteria usually stops here. The patient gradually quiets down, the movements become less and less assertive, and a crying fit or a sudden scream may terminate the affair. After the attack there is usually a condition of great fatigue and depression. Post-paroxysmal mutism is a frequent sequel, and mild aphasic or paraphasic disturbances are frequent. Voss reports the finding of a myasthenic reaction.

A neurological examination during a quiet stuporous interval, or post-paroxysmal stupor, usually reveals no marked changes. The pupils are usually widely dilated and react to light for the most part; Voss and others report slow reactions; loss of pupillary light reaction is known; Redlich's studies show considerable variability; sensory tests are obviously impossible, analgesia being usual. Signs of organic paralysis are absent, and the deep reflexes are not markedly altered, but the skin and mucous reflexes may show marked diminution. In rare instances only will one find the patient passing urine and fecal matter.

*Postconvulsive Period.—Dream States.*—The multiplicity of paths along which the major hysterics will travel is enormous, but about one-half of them, according to Pitres, less according to Binswanger's and Voss's observations, will pass into a condition of lethargy, in which they may remain for hours, days, in some instances, weeks. These are the patients concerning whom one reads so much in the yellow press—the "living dead" and "buried alive" cases.

These patients lie with closed eyes, immobile. The respiratory movements are hardly appreciable, and they often go for days without

attending to the wants of nature. Careful examinations shows that the urine leaks away little by little in some patients; passing the feces in bed is also reported. In others there is a condition of more active delirium; they talk, coherently or incoherently, to imaginary personages about them, a sort of semi-dream, spoken aloud, abounding in phantasy, and showing on analysis much of the characteristics of a dream, at times a typical delirium. Some patients in hysterical dream states show almost all of the characteristics of a katatonic dementia præcox, while, again, others behave like mildly confused manic-depressives.

The varieties of mental disturbance which may be arbitrarily arranged in this third group are countless. The attack often serves as the point of departure for many of the interparoxysmal phenomena. The patients wake up, lost, as it were, to their surroundings. They may have gone away from home in a somnambulistic state, and have little memory for what has happened, or only very hazy or isolated bits of memory concerning their actions. Patient analysis may bridge over the gaps and reconstruct a history of the mental wanderings, but unless the most severe self-critique is invoked, many of these attempts will reflect the subjective romantic dispositions of the investigator more often than they do the memories of the investigated.

It was a common experiment in the Paris hospitals in the days of Charcot, to press upon the ovarian region, mammary region, etc., to produce such major hysterical attacks, and as a result an extensive literature collected regarding these so-called hysterogenic zones; Charcot had his patients so trained that pressure on an ovary would cause an attack, pressure of the mamma would bring it to a close, and pressure on the two areas at the same time would cause no reaction. All this was nothing more than pure training. Scientifically one calls it suggestion; a great deal of it was humbug; not a malicious sort of deception, but a type of superabundant, uncritical faith shared in by both physician and patients. Hysterogenic zones there are none, if one restricts the meaning to the direct influence of definite peripheral irritations as capable of including hysterical attacks without the influence of psychological, suggestive factors on the part of the investigator. Again, it has been denied that major hysterical attacks could be consciously induced by the mere willing, but there is evidence that negatives this view.

**Course and Length.**—No two major hysterical attacks are alike. In general they are prolonged, usually lasting an hour or so, occasionally days, while lethargic states are known to persist several weeks. Some attacks are as short as a minute or two. Status hystericus lasting for several days is described.

**Varieties of Attacks.**—Kaleidoscopic varieties are to be expected. Monophase attacks are seen in those that show only the contortions, or others who fall directly into a lethargy or narcolepsy; more rarely mono-epileptoid phases are observed, and in these differentiation is difficult. Isolated cataleptic phases were frequent in the earlier descriptions; most of these are now relegated to the katatonias of the dementia præcox group, although apparently true hysterical catalepsies with *flexibilitas cerea* and hallucinatory ecstatic postures are known.

**Rhythmic Movements.**—*Tremors.*—The full description of the innumerable varieties of tremor in patients with complete or fragmentary hysterical reactions is due to the labors of the Charcot school. The three-fold classification that they offer (*a*) trepidation, (*b*) vibratory, and (*c*) intentional tremors has the merit of simplicity even if incomplete. Simple tremor when the hands are at rest is extremely frequent. Static tremor, when the fingers are strongly extended and spread apart, is less often seen, while intention tremor is comparatively rare. Mixed forms are the rule, and the polymorphic character of the tremor is diagnostic. All these types of tremor are usually rhythmic, but irregular, ataxic, choreic-like tremors are observed. The localization is manifold, eyelids (Rosenbach), shoulders, fingers, legs, tongue, mouth, etc. When involving the muscles of the mouth or tongue one obtains the characteristic stuttering, or irregular pseudo-paralytic speech disturbances. Horizontal axis tremors of the head may resemble early paralysis agitans tremor.

In the hands they may be unilateral or bilateral, local or general, quick, vibratory tremors—8 to 12 per second, slower, 5 to 12 per second (pseudo-mercurial), or even slower, 4 to 5 (paralysis agitans like). From another point of view the tremors may be peripheral, radicular, or spino-segmental. The chief diagnostic feature of the hand tremors is that they rarely conform to a single type; they are polymorphous. Hysterical tremors are absent during sleep. Attention directed to these hand tremors and attempts at restraining them usually result in their increase, and sudden emotional shock usually increases them greatly. In the act of eating they usually diminish or disappear. Isolated attacks of tremor are of further interest; such may persist for a week or so.

*Intention tremor* undoubtedly takes place in hysterical affections. It may resemble that of multiple sclerosis very closely, and is not infrequently met with in traumatic cases, especially in litigated ones. The combination of absent tremor in the supported hanging hand with intention tremor in hysteria is seldom met with. It is not to be forgotten that true multiple sclerosis tremor is all too frequently diagnosed as hysterical. The coincidence in similar localization of tremor with changes in sensibility is diagnostically especially significant.

Further rhythmic motor phenomena are hysterical hiccough, hysterical coughing, often extremely confusing, abdominal spasms, and hysterical shaking (shakers). Hysterical stuttering is a particularly obstinate type. Hysterical asthma, spastic hysterical aphonia (laryngismus hystericus) hysterical tachypnoea, dyspnoea, all fall within the rhythmic affections of the respiratory apparatus. Just which of these may be attributable to the influence of suggestion and which fall in the group of faulty diagnosis in Babinski's dismemberment time will decide.

*Coördinated Impulsive Movements.*—Hysterical crying and laughing are among the most ordinary explosions of hysteria, occurring for the most part in young women about the time of puberty. The middle grades of the hysterical temperament are prone to this type of expression, and in the histories of most hysterics one reads that such emotional expression was common in the earliest phases.

Binswanger offers certain diagnostic and prognostic signs concerning

these attacks and the severity of the hysterical neuropsychosis in general. In one class one observes them after emotional shock in the absence of other factors. They are then to be considered as the expression of a pathologically emotional irritability, and the explosion is out of all proportion to the affect shock both with reference to its intensity and its duration. They betoken constitutional inferiority, usually on a definitely hereditary basis, particularly of the same type of disturbance in the ascendants. Binſwanger is inclined to regard them, when symptomatic, of less grave importance than those of a second group in which an acquired or congenital neurasthenia lays the foundation. In the former group they represent perhaps the only disease feature of the personality, while in the second they seem to indicate a general instability and are more frequently accompanied by a more complete hysterical symptomatology.

Other types of coördinated impulsive movement attacks are described, a number of which are characterized by great uniformity; grimacing, hopping, etc. The trend of modern investigation is to ally many of these cases with the milder and even severer dementia præcox reactions. A characteristic failure of many of the earlier investigations of these movements was in not keeping the patients under observation for a sufficient number of years. Had this been done the development of dementia præcox would have been recognized. The symptomatology of hysteria might have been poorer, but a more correct understanding of the process would have been gained.

**Arrhythmic Movements.**—*Choreiform.*—While the tremors are usually rhythmical, irregular, incoördinated, unwilling, and large, movements of the choreic type are occasionally met with, which offer particularly difficult diagnostic problems. This is all the more striking since combinations of hysteria with chorea are observed. Imitative choreas, which may attain epidemic proportions, are matters of history. Charcot complicated the whole question by assuming the essential relationship of chorea and hysteria, a view that has support only on superficial grounds.

In all choreiform affections the important factors of infection or excessive growth with mental or physical fatigue should not be overlooked. To call a chorea hysterical in the absence of rheumatic fever is nonsense. Hysterical hemichorea is known. All patients with chorea are entitled to a rigid neurological examination, especial stress being laid upon the reflexes, and the more recent symptoms of involvement of the cerebellar and cerebrospinal systems. Spinal puncture should not be omitted in the severe cases. Chorea hysterica in the face of rigid neurological and psychiatric examination is becoming rarer and rarer.

*Myoclonus.*—Here the movements are larger, involving whole, functionally related muscular groups, and the contractions are rapid, tic-like in character. Whether there is a hysterical myoclonus is as difficult to decide as to assume practically all myoclonic reactions to be hysterical. To refer them to the tics is simply to push the psychogenic factor farther back in the past. This entire group of affections needs revision.

*Tetany.*—The difference between a true tetany and hysterical tetany reaction is difficult to state. The presence of Chvostek's, Erb's, and Trousseau's phenomena, with the special factors of occupation and of

etiology, go far to establish the diagnosis of essential tetany. The test of calcium therapy affords further information. From the clinical point of view the signs known are only generally indicative; they are not positive, not even Erb's phenomena. In many hystericals electrical hyperexcitability of the muscles is marked.

**Akinetic Motor Phenomena.**—*Paralyses.*—Weakness or loss of muscular power is among the most frequent of the more marked manifestations. There is the greatest variety, with reference both to extent and grade of akinesis. For many years clinical neurology was without definite criteria of the psychic nature of these pareses and paralyses, and it has been only within recent times that the essential differences between organic and functional paralyses have been definitely formulated. Statistically considered, about 25 per cent. of hysterics show definite akineses. In Briquet's series there were 125 in 405. Landouzy reports 40 paralyses in 370, while Pitres' figures are less than 17 in 100 with paralyses of the extremities. Binswanger reports 4 in 109, remarking that the paralyses are less frequently seen in Germany than in France. Voss, working largely with Russians, finds nearly as many as did Briquet, 27 in 123. Percentages will depend largely upon the type of clientele, and the special suggestive factors. Personal figures drawn from three types of clientele show less than 5 per cent. in what might be called private practice, perhaps 10 per cent. in hospital practice, and over 60 per cent. in litigation railroad accident work. Statistics are largely fortuitous.

Muscular weakness, myasthenia, of varying grades is found in the majority of hysterics; it may remain without developing farther, or be but the beginning of a paralysis. Maladroit suggestions can convert a paresis into a paralysis with surprising facility. These myasthenias, which are purely of psychogenic origin, due to the sense perhaps of helplessness, of anxiety, of dread, fear, displeasure, pain, etc., which affect influenced psychical processes and exert a depressing effect upon the motor innervation, are to be distinguished from the neurasthenic myasthenias. Clinically considered, however, they are mostly hysteroneurasthenic in character. Difficulties in diagnosis from organic myasthenias are often extreme, especially in early multiple sclerosis and certain obscure spinal-cord lesions, small circumscribed myelitis, slowly developing tumors, low-grade pressure neuritis, certain intoxications, etc.

Of the most outspoken paralyses, hemiplegia, paraplegia and monoplegia are the more striking; mixed types, are also met with.

*Hemiplegia.*—This occurs in about one-half of the total paralyses (Ziehen). It may be incomplete—diplegic, arm and leg without cranial nerve involvement—the commoner form—or rarely with cranial nerve additions, or mixed hemiplegic with the opposite arm or leg. Still more rare are the quadriplegias, and alternate hemiplegias are curiosities. The onset of the hemiplegia is nearly always acute, often accompanied by a sense of weakness or giddiness, and almost invariably accompanying an affect shock; many develop after a major hysterical attack. The paralysis is at once manifest, and only rarely advances slowly. Occasionally pain, nausea, and vomiting accompany the paralysis.

Clinically considered, these paralyses lack the signs of organic disease.

The absence of involvement of the cerebral neurone is demonstrable by the absence of pathologically increased tendon reflexes, clonus, increased tonus, of Babinski's phenomena, of Oppenheim's reflex, or modification of Schaffer's paradoxical reflexes, of Van Gehuchten's inguinal cutaneous reflex, the femoral reflexes of Remak, of Grasset's, Peaucier's, Hoover's, and Babinski's hip-thigh, associated movement signs, etc. The integrity of the peripheral neurone is evidenced by the presence of knee-jerks and of the Achilles jerk, the absence of atrophy (until contractures may develop after long disuse), no nerve tenderness (unless psychogenic), or unaltered electrical excitability, hypotonus, trophic disturbances, etc.

These are the usual diagnostic signs separating functional from organic cases, yet a review of recent literature emphasizes the need of excessive caution in attaching too much or too little importance to variations which undoubtedly exist. Thus while it is clearly realized that the tendon reflexes should be normal, Babinski maintains always, the knee-jerks are usually very active. Ankle clonus, called by some pseudo-clonus, has been reported by Binswanger, Sternberg, Westphal, and Voss. Oppenheim describes foot tremor, and Dejérine has demonstrated that patellar and foot clonus are exceptionally present in hysterical cases. Of recent years considerable discussion has centred about the possibility of the presence of inferior cutaneous reflexes, particularly Babinski's phenomena in hysteria, heretofore considered absolutely diagnostic of organic involvement of the pyramidal tracts. Roth, Van Gehuchten, and others, however, report the occasional presence of a Babinski toe phenomenon. Since modern conceptions of the many factors which may cause a Babinski toe phenomenon have been so amplified, definite judgment concerning this is premature.

Involvement of the motor cranial nerves seldom occurs in hysterical palsy. The cases reported of oculomotor and abducens paralysis are extremely doubtful (Borel, Parinaud), and the differentiation between a paresis and a spasm is extremely difficult. The cases of ophthalmoplegia externa should also be judged with the idea of hysterical contractures in view. Hysterical ptosis is not infrequent.

*Facial palsy* is of particular interest as a hysterical sign. All three branches may be involved, although participation of the supra-orbital branches is exceedingly rare. Special attention must be directed clinically to the danger of seeing a palsy on one side when there is in reality a hysterical contracture on the opposite side.

*Glossolabial* involvement is reported (Remak), and considerable variation exists regarding the position of the tongue and the palate. Most of the anomalous cases seem to consist of a mixture of spasm and of paresis. In the hysterical aphonias the movements of the palate have been observed to be less ample. Inferior alternate paralyses with crossed facial are reported. They need to be carefully weighed (Voss).

Laryngeal palsy may also be reckoned among the paralyses, more frequently bilateral than unilateral, it is rarely complete. Laryngoscopically, the picture is usually negative.

*Paraplegia*.—Paraplegia superior is extremely rare, while paraplegia of the lower extremities is more frequent, although statistics concerning

its occurrence are not available. Inferior paraplegia is found either as a typical paralysis of the lower extremities, with inability to move the limbs, complete or in part, or as an abasia (*astasia-abasia*), in which there is conservation of the power to move the limbs while in a horizontal position, but inability to use them in the vertical position. Both the flaccid and spasmodic types of paraplegias have their analogues in the hysterical paraplegias, and contractures with these paraplegias are comparatively often met with. Transitional types are the rule.

Trauma, especially in young girls, or even children, is the most frequent exciting cause. Railway accident hysterical paraplegias are of comparatively frequent occurrence. The onset is, as a rule, brusque, although its gradual development is observed, and Voss states that excessive tire of the lower extremities in anemic young girls may be considered as a contributory cause. The distribution is apt to be extensive, although restrictive movements may be preserved. The patellar reflex is apt to be exaggerated in both types, but the absence of the signs mentioned in the discussion on hemiplegia is indicative of the psychogenic nature of the disturbance. The functions of the bladder and rectum are usually normal, although several authors note involuntary defecation and urination. Mistakes in diagnosis, or imperfect observation, are here to be particularly guarded against, since the symptoms from small spinal-cord hemorrhages are not infrequently diagnosed as hysterical.

The distribution of the anesthesia will largely depend upon the method of examination. A maladroit examiner can find almost anything he wishes, and since the usual methods for examining for sensory disturbances are largely tinctured with clumsy suggestions, the beautiful figures of the Charcot School students are better evidences of such methods than scientific documents. In examining for anesthesia, the real point should always be guarded. The patient should never be asked if she feels, but should be questioned to distinguish between kinds of sensations. A consistent anesthesia to all the types of possible test has never been encountered. The classical representations show a sensory distribution practically coinciding with the extent of the motor loss, but a great variety of irregular distributions has been figured.

Atrophy may follow contracture, but without reaction of degeneration, although Dubois has well shown that a diminution of both galvanic and faradic irritability in the majority of cases is possible. After years of inactivity, paraplegic contractures may be absent.

*Astasia-abasia*.—This is an irregular incomplete type of paraplegic disturbance, first described by Blocq, in 1888, differing from true paraplegia briefly in that there is no paralysis or ataxia of the limbs in the sitting or reclining position, but only when the patient tries to stand or to walk. It is undoubtedly of psychogenetic origin, and, hence, should be considered as a hysterical complication of the other psychoneuroses, or an anxiety neurosis, neurasthenia, etc.

*Pseudo-tabes*.—Pitres has given a full analysis of this allied disturbance. Pains, incoördinated ataxic movements, disturbance of the eyes, analgesia, etc., make a differential diagnosis extremely difficult at times. It is an interesting fact that Charcot made a diagnosis of true tabes in a patient



who went to Lourdes and recovered. Hysterical symptoms in a tabetic are frequent, but the diagnosis should not be difficult.

**Monoplegia.**—Hysterical monoplegias are extremely frequent, and very irregularly distributed. Mention has been made of the paralyses of the third, fourth, or sixth nerves, of the facial and of the muscles of the pharynx and larynx. Even individual muscles of a nerve innervation group may be involved as in hysterical ptosis, single, or double, spastic or flaccid, internal or external rectus, etc., with nystagmus. Brachial monoplegias are perhaps the most frequent, leg monoplegias are rarer.

**Disturbances of Sensation.**—A searching study of hysterical anesthesia is lacking. Believing it to be premature to accept *in toto* the dictum of Babinski, that there is no such thing as hysterical anesthesia, a brief recapitulation of the knowledge concerning these phenomena is desirable.<sup>1</sup>

**Disturbances of Touch.**—These consist of anesthesia, hypesthesia, and hyperesthesia. Anesthesia may be general, unilateral, isolated in spots or patches, or involving symmetrical regional areas. Hypesthesia, *i. e.*, incomplete anesthesia, is much commoner than anesthesia. Total general anesthesia is probably extremely rare. Pitres' statement that 20 per cent. of the cases show it, is one of the extravagances of the Charcot period. Binswanger considered only six of the many reported cases to be valid, and Voss added six from the literature as probably authentic, and two of his own observation. Some of these patients showed "dream" state conditions, and it is difficult to estimate their value.

Hemi-anesthesia or hemi-hypesthesia is not infrequent, although the usually accepted teachings of Charcot are certainly defective. The left side appears to be much more often involved, and considerable variation in intensity exists in the different portions of the affected side. Crossed anesthesia, *i. e.*, face of one side, and body of opposite side, is also reported. Bizarre groupings, such as involvement above the waist on one side, and below the waist on the opposite side, are known. It is not impossible that some of the crossed anesthetics have been really due to minute thalamic lesions. Patch-like anesthesia or hypesthesia is common. The breast and forearm are especially involved, but the irregularity in position, form, size, constancy, etc., is bewildering.

Geometrical distribution, the well-known stocking-and-glove type, which rarely corresponds to any anatomical nerve area, is classic. The border of the anesthetic area is apt to be very indefinite, not only during an examination, but particularly on comparison with preceding examinations. The patch-like anesthetics show a similar, if not greater, variability. As a rule, the patient is unaware of the anesthesia, and in but few cases does it cause any inconvenience. Other patients complain of pain (anesthesia dolorosa) or pricking, or compare the sensation to the crawling of ants, or the feeling of a limb that has gone to sleep.

Anesthesia develops suddenly, as a rule, either following a major attack, or without such a precedent. It would appear that few old hysterics are free from anesthetic concomitants. The increasing opportunity for suggestive action is here manifest.

<sup>1</sup> See Voss (Schmidt's *Jahrbucher*, February, 1909) for a critical summary of Babinski's teachings concerning anesthesia.

*Temperature Sense.*—Loss of ability to distinguish heat and cold appears to follow the modification of tactile anesthesia, but cold nearly always has a striking effect upon the hysteric, especially when applied *en masse*, as by a shower bath or cold pack. A few, not truly convincing, cases of dissociation of temperature and tactile sense are on record, also cases of dissociation of warm and cold. Patients with loss of temperature sense rarely burn themselves, although burning in hysterical thermanesthesia is recorded. There are no complete studies in accord with Head's teachings regarding epicritic and protopathic sensibility.

*Deep Sensibility.*—In a few isolated instances deep sensibility seems to be affected, but it does not necessarily impose its pathological corollaries—ataxia, etc. Hypesthesias and hyperalgesias are more frequent than deep anesthetics. Dissociations with intact tactile and lost deep sensibility, or vice versa, are among the hysterical anomalies recorded that require more corroboration and more exact observation.

*Mucous Membrane.*—The sensibility of the mucous membranes may be modified in a manner quite analogous to that already noted for the skin. Thus anesthetics of the mucous membrane of the mouth, nose, urethra, rectum, ear-drum, epiglottis, etc., are recorded. Thaon makes the statement that in one-sixth of all hysterics there is anesthesia of the epiglottis, yet respiration pneumonia does not occur as a result.

*Pain Sensibility.*—Analgesia and hyperalgesia are both present. General diminution of pain sensibility, hypalgesia, is about as frequent as general hypesthesia, and is found under similar conditions. Absolute analgesia is found in a few hysterical dream ecstatic states. Hemi-analgesia and hemi-hypalgesia run *pari passu* with the hemi-anesthesia and hemi-hypesthesia, and crossed varieties are described. The usual pupillary response to painful stimuli is obtained in many of these cases, thus the integrity of the reflex mechanism is assured even if there may be repression of the psychical pain associations.

*Hyperalgesia* is a common hysterical symptom. In one form or another it enters into almost every hysterical history. Hysterical points or zones of tenderness on deep pressure, usually unilateral, are extremely frequent. The most common of these are the iliacal, or so-called ovarian, pressure point, also present in men, inguinal pressure point, just above Poupart's ligament, epigastric point, mammary point, jugular point, the supra-orbital and infra-orbital points, and the vertebral points. There are extended discussions of the location of these points and their relation to the so-called hysterogenic zones. Suggestion is the important element, but there are certain areas where pressure with suggestion seems to have more influence than other zones; the same phenomena may be brought about, however, by other suggestive means. After all, the point has no real relation to the disease. The most classical points are usually those upon pressure of which physiological stimuli are more readily set up in the normal individual. Pressure upon other zones, or even the same point, may bring about the cessation of an attack. Pressure on these points may also determine localized pains, paralyses, contractures, ataxias, etc., and vice versa, it can be used to cause these symptoms to disappear.

*Spontaneous Pains*, found in perhaps 80 per cent. of all hysterics, are exceedingly variable, but may be roughly classed as follows:

(a) *Topalgias* or circumscribed painful areas having no anatomical relation to any known nerve distribution. The study of these has been much confused by lack of careful consideration of Head's zones of reflex visceral disturbance and vice versa, hysterical topalgias have been often considered as evidence of visceral disease. The occurrence of topalgias as isolated phenomena should be interpreted more as evidence of reflex visceral disturbance than as indicative of hysteria, and the modern tendency is to recognize more and more the strength of Head's position, that a great mass of symptoms which have been regarded as hysterical are in reality but the peripheral reflex disturbances due to visceral disorder or disease. Further, many topalgias are found in the neurasthenic.

(b) *Neuralgias*.—In the earlier studies these were described as occurring frequently, but more careful inquiry has revealed faulty diagnosis, so that, at present, a neuralgia, even when apparently not conforming to the known anatomical distribution of a nerve trunk, should be diagnosed as hysterical only after careful elimination of organic features. Examination may reveal a cervical rib to account for a persistent brachial neuralgia or sugar in the urine may reveal the diabetic origin of another.

(c) *Arthralgia*.—Pains in the joints offer particularly difficult problems. Hysterical coxalgia should never be so diagnosed in the absence of a radiographic examination. Since the advent of such methods they have been much less frequently diagnosed, even when the arthralgias are accompanied by hypesthesia or hyperesthesia, contractures and disturbances of gait, apparently hysterical. Certain cases are so quickly relieved by suggestive therapeutics that the occurrence of true hysterical arthralgias seems certain.

(d) *Enteralgia*.—Many striking cases of visceral pains of undoubted psychical origin are in the literature. Mention has been made of hysterical appendicitis, concerning which A. Dubois has written an extensive monograph. Hysterical ileus, with pain and fecal vomiting, is held by Babinski to belong to the group of "simulation," in which opinion Strümpell shares, but Bergman has collected the older cases, Voss the newer ones, and Naunyn believes it a possible hysterical condition. Studies on antiperistalsis by Muhsam<sup>1</sup> seems to point to the possible psychogenic origin of such reversed peristalsis. Vomiting is a frequent hysterical symptom, and even the hyperemesis gravidarum is held by E. H. Müller to be of possibly psychical origin.

(e) *Cardialgias*.—Binswanger described cardialgia as a frequent hysterical symptom. In its severest forms—angina pectoris hysterica—it is undoubtedly very rare, but as palpitation, sense of oppression, irregular pains, quick pulse, dropped beats, etc., it is frequent. Hysterical bradycardia has been described by Triboulet.

In addition to the false cardiopathies of both hysterical and neurasthenic origin, there are the false gastropathies, mental anorexias, false enteropathies, false genitopathies, etc., on which Dejerine has cast such

<sup>1</sup> *Mitt. a. d. Grenzgebiete d. Med. u. Chir.*, 1900, vi.

a wholesome illumination. They have often been indiscriminately herded with the hysterical symptomatology, or with hysteroneurasthenia; many are found in Janet's psychasthenic group. In their purest expression they should be kept in a group by themselves as the false pathies. They frequently are complications of hysteria, and are too largely the product of bad medical suggestion.

*Disturbance of Vestibular Apparatus.*—Dizziness of vestibular origin as a pure hysterical reaction is undescribed, although Frankl-Hochwart's and Bouyer's cases of pseudo-Ménière's disease have been referred to this category, and hysterical aural vertigo has been described. Voss has also given an apparently clear case of aural vertigo of hysterical origin, operated upon without finding any organic cause. A close reading of these cases shows many deficiencies of examination sufficient to exclude them from the category of well-known but transient or even mild organic vestibular affections. It is better perhaps to acknowledge them as due to somatic causes yet unknown than to shut ones eyes to careful methods of examination by calling such hysterical. The possibility of the occurrence of the hysterical reaction due to cerebellar (vestibular) disturbance is not to be overlooked. The same is true for a large number of the dizzy spells frequently met with. Binswanger has correctly estimated these as common complaints arising from a variety of transitory causes, weakness, tire, fatigue from eye movements, gastro-enteric disturbance, etc.

*Smell.*—The percentage of normal variation in smell is not yet definitely established, hence the work of Lichwitz, who found complete and half-sided anosmias and other anomalies of smell, is not conclusive. Irregularities of smell in the non-hysterical are very commonly found by Ziehen, who makes a systematic investigation of smell in every case in his clinic. Gilles de la Tourette is inclined to regard such disturbances as of diagnostic value. Babinski fails to find any marked anomalies unless the methods of investigation are palpably suggestive. Voss finds 30 per cent. of his cases with anomalies. This is about the average run of variations found in the psychiatric clinic at Berlin for all cases. Voss finds the left side more often involved.

*Taste.*—Variations in taste for sweet, sour, bitter are not frequent, but are recorded by numerous authors. Hyperagusia is especially pronounced. Agusia is a frequent accompaniment of the facial anesthesia. Hemilateral agusia and hyperagusia are described.

*Hearing.*—Hysterical deafness is rare, yet recorded by Binswanger Habermann, Magnus, Barth, Walton, and others, yet changes in modulation of the voice, so characteristic of organic affections of the ear, are practically absent. Weber makes some observations to the contrary which are striking. Bone conduction is also modified. Voss has described double hysterical deafness in combination with hysterical paraplegia.

*Sight.*—Double-sided blindness is rare, and few reported cases are free from scepticism. Unilateral blindness is frequent. Normal eye grounds, unmodified pupillary reactions, acute onset under psychical shock are the usual diagnostic features, taken in connection with the other features of the hysterical personality. The test with colored glasses and colored word stimuli, with later association tests, reveals the fact that the

patients see, and the image is not in the subconscious. It is voluntarily repressed. Freud's studies on psychogenic blindness are valuable.<sup>1</sup>

The lid reflex to sudden illumination is a valuable diagnostic sign. The consensual light reaction has not been carefully observed. Voss reports a case with its diminution in a unilateral amaurosis.

Diminution of vision as a subjective complaint is extremely common; it usually involves the left eye, while restriction of the visual fields has become a classic since the work of Féré. Babinski denies its presence absolutely, and claims it to be the product of *maladroit* suggestion. Such has been largely the writer's experience. Such limitations are described as concentric and bilateral, and are claimed for a third of all the cases; Voss gives 25 per cent. Increased fatigue, inability to concentrate the attention, etc., are among the etiological factors. It is subject to great variation, and simulation and exaggerations are frequent.

Central scotomata, sector and ring-shaped defects, are among the rarer anomalies on record. Disturbances in color perception are also on record. Achromatopsia for single colors is the most frequent, although apparent true color blindness is described. Seeing double with one eye (*polyopia monocularis*) was first described by Parinaud. Within recent years Voss reports four cases and negatives the ciliary accommodation cramp hypothesis in favor of its being dependent on an amaurosis. Ziehen suggests a hallucinatory basis, especially for Ulrich's patient, who saw things six times with one eye.

Pupillary disturbances may originate from psychical influences. Pupillary immobility is claimed as a hysterical symptom, but most authors maintain that even in the severest hysterical attacks absolute rigidity is absent. Voss believes he has seen slowed reactions during an attack—attention has also been directed to this author's observation regarding unequal pupillary reactions in one-sided amaurosis. Spiller has reported irregular pupillary phenomena in a case of hysterical chorea. Micropsia and macropsia are rare phenomena (Janet, O. Fischer).

**Vasomotor and Trophic Disturbances.**—Here may be grouped a number of phenomena which have excited great discussion; hysterical fever, changes in the skin, oedema, urticaria, with modifications of the secretions, and metabolic disturbances serving as the chief examples.

*Hysterical Fever.*—That hysteria may, without other complicating factors, give rise to hyperthermia is the conclusion of Manahiloff, who, in a *Montpellier Thesis* of 1903, has gone over the entire field critically and thoroughly. The fever is due to a disturbance of the heat regulatory centres. Kausch,<sup>2</sup> in a collected criticism, comes to the same conclusion. There are not wanting, however, those who contest the validity of a hysterical fever. Oppenheim believes in its possibility, and has recently reported personal observations, and in Voss' latest monograph the evidence is confirmatory.

The temperature curve is not characteristic, and the fever is found usually only in the severest forms, especially those with major convulsive phenomena. The grade of rise is not great, as a rule, yet temperatures

<sup>1</sup> *Samml. klin. Schriften*, iii; see also Ames, *Psychoanalytic Rev.*, 1913, i.

<sup>2</sup> *Mitt. u. d. Grenz. der Med. u. Chir.*, 1906.

of 104° to 106° F. have been recorded. In certain cases with fever the accompanying pulse and respiratory changes have not taken place, while in others, dyspnœa and tachypnœa occur. Curious anomalies are reported by several observers, such as an axillary temperature 2.3° F. higher than the rectal; differences on the two sides of the body, etc. Such variations are known to exist in organic lesions of the nervous system. A diagnosis of hysterical fever should be made only after every possible cause of hyperthermia had been excluded. A diagnosis of hysteria founded on fever alone is untenable.

*Skin Changes.*—Notwithstanding the excellent review of Cassirer<sup>1</sup> the question of the occurrence of hysterical lesions of the skin has not reached a unanimous conclusion. Babinski classes the usual skin eruptions, bullæ, gangrene, ulcers, etc., among the deceptions or artefacts. There is no doubt that many are such, and there is further evidence to support the contention that hysteria may be solely a complicating factor in an individual with peculiar trophic skin phenomena, but there seems no reason for doubting the psychogenic origin of many skin lesions.

Among the contributions to the literature of multiple gangrene of the skin thought to be of hysterical origin are those of Kreibich and Matzenauer, who observed markedly increased vasomotor irritability in hystericals, in some of whom the slightest skin irritation would lead to gangrenous necrotic ulceration. Bettmann has reported a case of gangrene in a hysterical individual, due to the application of an extremely weak lysol solution, and in many hystericals the skin may be excessively delicate in its reaction to external irritants. It is not a far cry to interpret an acceleration of a reparative process to the same hysterical type. At any rate, in spite of the practically unscientific aspect of the Lourdes "marvels," certain interesting rapid cures of ulcerous processes are worthy of more careful consideration than has hitherto been given them.

Bullous dermatitis is not infrequently observed as a hysterical phenomenon, while among the rarer types of so-called hysterical skin disorders are reckoned erythema, urticaria, dermatographia, ecchymoses, chromidrosis, vicarious bleedings, etc. They are to be accepted with considerable reserve for the vast majority of these skin changes have been artefacts. Dupré has suggested the term *mythomania* to cover a number of these chronic self-mutilators. Yet there remain a few cases in which fraud cannot be proved. Certain cases of circumaural acne, of psoriasis and of urticaria offer examples of auto-erotic hysterical conversions.

*Œdema.*—The instructive discussion of the Paris Neurological Society did not settle the debated point of hysterical œdema, which was first noted by Sydenham, nor could any unanimity be reached concerning the relation of suggestion to œdema. The cases reported have been many, and a general classification into white and blue œdema is made, the former being soft, the latter hard. These come on rapidly, usually following an affect shock, and disappear as quickly. Pitres has made them disappear apparently under the influence of a magnet. They may be associated with sensory or motor symptoms and be distributed as a

<sup>1</sup> *Die vasomotorisch-tropischen Neurosen*, 2d edition, Berlin, 1913.

hemiplegia, a paraplegia, or a monoplegia. In some instances of blue oedema the skin is infiltrated and pits on pressure. In other cases there seems to be a local vasomotor paralysis, with no infiltration and no pitting. Andemach<sup>1</sup> has reported an unusually well-authenticated case, also with loss of the knee-jerks. The exclusively hysterical nature of these oedemas is not yet conclusively established; but the preponderance of evidence is in favor of the possibility of their psychogenic origin.

**Bleeding.**—Hysterical phthisis has been, for the most part, relegated to the mistakes in diagnosis, as well as practically all of the gastric and kidney hemorrhages, but a number of observations of bleeding from the skin, mucous membranes, or viscera, often termed vicarious menstruation, etc., remain, after the deceptions are exhausted, as bona fide hysterical phenomena. Many of these patients show a number of related vasomotor disturbances. The bleeding may be scanty or profuse, in which latter case, however, the general health does not seem affected.

**Secretory Disturbances.**—The influence of hysterical emotional states on the secretory activities is evident, and a number of anomalies are recorded. The sweat glands usually functionate normally, but lack of perspiration, which may be unilateral, has been observed. Local hyperidrosis has also been described. Under artificial conditions, such as medication with pilocarpine, the sweat secretion has been made symmetrical, even in a hysterical individual who was evidencing unilateral secretory anomalies. The salivary secretion is more often diminished than increased. Yet a striking case of sialorrhœa has been described by Matthieu and Roux, in which there was a secretion of two-thirds of a liter daily. This case was complicated by excessive vomiting. Increased flow of tears or absolute dryness may be met with. Increased bronchial secretion, accompanied by coughing; or excessive swallowing of laryngeal or pharyngeal mucus, may also be noted, as well as increased nasal secretion with sneezing attacks. From the diagnostic side it may be borne in mind that unilateral and even bilateral disturbances of these secretions diagnosed as hysteria have in some cases developed multiple sclerosis, or syringomyelia.

The urinary secretion is frequently very variable; at the present time one meets with fewer cases of absolute anuria than in the Charcot days, especially since ureteral catheterization has been made possible, yet in a few instances one finds relative anuria in association with hyperidrosis or excessive stools. Simulation is the usual explanation, for Babinski the sole explanation, but there are definite cases of relative anuria. Polyuria is more frequent. Matthieu has reported a case in which 30 to 40 liters of urine a day were excreted. During major attacks an increase of urine may be expected. Incontinence is rare, but occasionally occurs either as a paroxysmal or interparoxysmal phenomena. In hysterical trance or ecstatic states attention must be directed to the bladder in order to avoid overdistension.

The intestinal secretions may be profoundly modified by the mental state. Cannon has shown for the intestines what his predecessors have

<sup>1</sup> *Münch. med. Woch.*, October 26, 1909, p. 2222.

shown for gastric motility and secretory activity. Diminished or increased gastric and intestinal secretions are among the most frequent anomalies in the hysterical. Hysterical diarrhoea is not infrequent, while hysterical constipation is very common, especially in anal erotic conversions.

**Metabolic Disturbances.**—The loss of flesh in some hysterics is classic. They may be true hysterics or the mental anorexias of Dejerine. A notable example of the "human skeleton" cured in recent years at Lourdes belongs to this group. The loss of flesh is usually the result of not eating, but some patients grow thin in spite of fairly normal feeding. A few cases of hysterical adiposis are recorded, but their exact significance is doubtful. The researches of Gilles de la Tourette have shown that in the interparoxysmal period there are no marked deviations from the normal metabolism. During the attacks, however, a modification of the usual relationship of calcium and magnesium to the phosphates of potassium and sodium may take place, *i. e.*, there may be an inversion of the formula of the phosphates. The most recent studies carried on in Binswanger's clinic have yielded no striking conclusions.

**Blood.**—No generalizations of value have yet come from blood studies. A recent work of Schultz shows that the oft-noted anemia may be associated with even an increased hemoglobin content; that the red blood cells are usually normal, and further, that as a result of the vasomotor disturbances in hysteria it is necessary to take blood for examination from different parts of the body. The conflicting results of previous studies are partly explicable because of lack of attention to this detail. The relationship of eosinophilia to the vagotonic constitution should be borne in mind in the study of certain patients.

**Reflexes.**—The corneal reflex, conjunctival and palpebral reflexes are frequently diminished. The corneal reflex is rarely absent, and the secretion of tears is not modified, as a rule, even in the presence of diminished corneal and palpebral reflexes. Irregular distribution in the sneezing reflex is occasional. It has little diagnostic significance, and loss of the faucial and pharyngeal reflexes, on which much stress has been laid, is of minor value. They are frequently very sluggish in the non-hysterical, especially those who have had much treatment of the throat. In the hysterical, irregularities in the faucial reflex are more apt to be present than in the non-hysterical. Little is known of the jaw-jerk.

The triceps and radial periosteal reflexes are usually increased. In the presence of contractures they may not be obtainable. At times they are lost, but their diagnostic value is minimal. Marked irregularity would point to organic lesions. The abdominal, epigastric, and cremasteric reflexes are subject to considerable normal variation, but when unequal, organic affection should be searched for; mistakes occur by the contraction of the abdominal muscles. Even in the presence of anesthetic skin areas the cremasteric reflex is rarely modified. The anal reflex may be absent; occasionally it shows hyperesthetic sensibility in much the same manner as one finds in vaginismus—also a rare hysterical reflex. Anal hypesthesia is not infrequent, especially in women, but what relation it has to the anal erotics of Freud is not yet determined.



The knee-jerks are usually increased, but, as a rule, symmetrically. Loss of the knee-jerk is undoubtedly present as a hysterical sign, although contested by Babinski. Many observations leave little doubt as to the actual occurrence of loss of knee-jerk during and after hysterical attacks. They are extremely rare, and may later permit of other interpretations. A similar position may be taken for the Achilles jerk. There is a known variability here. However, if tested by the kneeling method its absence is strongly indicative of organic lesion. In the observations of Nonne and Willroth, the interesting finding was a periodic loss of the tendon reflexes. In the interparoxysmal periods they were normal or increased.

**Psychoses.**—The use of the term is to a certain extent tautological, since from one point of view hysteria itself is primarily a mental disorder, but putting aside a too strict interpretation of the word psychosis, under this head can be grouped a series of phenomena that bear at times a close relation to the somatic disturbances.

The most striking of these profound mental alterations, and one which has been understood only within comparatively recent years, is the so-called hysterical dream state (*Dämmerzustand*). For years it has been known that following an epileptic attack or during an alcoholic debauch a patient may show a condition of dreamy delirium, which may not seriously interfere with his general orientation or conduct, but during which ordinary normal consciousness is not operative. Similar attacks are known to follow major hysterical seizures, or may originate without any convulsive antecedents. These are of shorter or longer duration, persisting from a few moments to perhaps a few weeks. Occasionally such a dream state may be terminated by convulsive seizures. The most striking of these states may be observed during major hysterical attacks, but there are other types during which the patients seem perfectly quiet and contained, or they are only dazed. If consciousness is deeply clouded, one finds these patients in a continuous slumber, with closed eyelids, sunken head, and relaxed muscles. They are the "living dead" frequently commented on by the lay press.

In another type one finds the patients about, even attending to their daily affairs, but doing them in a mechanical automatic manner, engaged, as it were, in a deep reverie all of the time. Patients in this condition have taken long voyages, and on coming to themselves are at a loss to account for their actions. Many fugues, or flights, are due to these hysterical attacks. Occasionally such people commit criminal acts, and important and difficult medicolegal problems arise. Somnambulistic performances belong to this general type of phenomena, although here the clouding of consciousness is rarely as profound as in the dream states following convulsive attacks. In certain of these dream states the patient rehearses in his delirium occurrences that produced the initial affect shock. Whether the dream state be accompanied by motor restlessness, agitation, or by stupor and lethargy, there is usually after the attack complete or partial retrograde amnesia. It is rarely a complete amnesia, but it may be.

A further feature of considerable importance is the occurrence of a

peculiar type of conduct which was first clearly set forth by Ganser, of Dresden, as "Vorbeireden." Many of the patients, especially the younger ones, show a peculiar foolish behavior quite like young boys of from eight to ten years of age. They are very short and snappy in their answers, burst out into uncontrollable laughter, imitate the cries of animals, and are extremely difficult to manage. In answer to questions they give indirect or inverse answers, half answers, contradict themselves—very obviously at times, apparently not caring just what they say. At first sight their answers seem like deliberate attempts at falsehood or simulation, but the great similarity in this type of case is so striking as to permit the recognition of the Ganser symptom. This type of answer is not peculiar to hysteria; it is found in alcoholism, in epileptic dream states, and is very common in dementia præcox.

Hysterical dream states may be confused with similar *dämmerzustände* of alcoholic, epileptic, or of traumatic origin. The diagnosis of a hysterical dream state is founded on the grounds of a dreamy disturbance of consciousness, suggestibility, an extreme changeability of the conduct, on foolish, childish conduct, and the presence of somatic hysterical signs. Diagnosis may be almost impossible in the presence of alcoholism.

It is of extreme medicolegal importance to recall that the motives of waking consciousness may be carried over into the dreamy state, and that criminal acts which seem to have a perfectly well-recognized motive may be performed in a state of consciousness in which the sick individual does not know the difference between right and wrong. The question of legal responsibility in hysteria is extremely knotty. It has rarely entered into the courts in the United States or England, but is well-recognized by Continental jurists.

Closely related to the Ganser syndrome is that of *pseudologia phantastica*, or pathological liars. This condition is well studied in the monographs of Binswanger and Voss, and particularly in the communication of Delbruch. Pathological hysterical swindlers have been made the subject of an important monographic treatment by Kraepelin.<sup>1</sup>

**Course and Prognosis.**—Ziehen has coined the word "delective" to express the multiplex combinations of the hysterical manifestations. By this is meant that any symptom in the broad group may be chosen by the individual, and that in successive attacks the picture may be quite dissimilar. This renders it almost impossible to describe the course of this psychical reaction. In general, it is a chronic affair, no consideration being given at this time to the purely symptomatic hysterical outbreaks which accompany other disorders.

It has been only recently that the suspicion has become a conviction that many of the symptoms which mark the course of hysteria are of medical manufacture, and the varied groupings of the picture come into existence in response to suggestions on the part of the attending physician, nurses, or anxious friends. Psychical contagion is one of the most difficult features to combat in limiting the symptomatology. Too great solicitude that the patient should not tire herself, or exercise too much,

<sup>1</sup> *Allg. Zeitsch. f. Psych.*, 1906.

may be the point of departure of a paresis or paralysis. This plays a great part in modifying the course and in varying its manifestations.

Although the course in ordinary hysteria may be regarded as chronic, much will depend upon the element of causation, the subsequent affect shocks, and naturally the factor of most importance, the mental foundation of the individual. In a young individual, subjected to a single affect shock, other things being equal, one expects a less protracted course with fewer variations than in an older individual subjected to the vicissitudes of constantly recurring causes for emotional disturbance.

The prognosis in general, so far as the somatic signs are concerned, is good, but here a general rule will not apply to the individual case. There are certain hysterical individuals who never recover. The hysterical constitution is so definitely established that, although the individual paralysis, analgesia, or anesthesia is recovered from, the patients go on to other manifestations. They become chronic invalids, the despair of all those connected with them. The severe degenerative hysterias—usually superior feeble-minded deviates, with hysterical complications, rarely ever recover. In the hysterical complications of organic disease one does not look for recovery. The neurasthenic hysterias present a hopeful prognosis if the affective incidents can be regulated and the struggle for existence better adjusted.

The traumatic hysterias, barring the other factors, *i. e.*, mental inferiority, psychopathic constitution, alcoholism, neurasthenia, etc., have a fairly good prognosis. The continuance of litigation, with its emotionally disturbing factors of expectancy, the desire to get even, and the constant suggestion to make one's self out as sick as possible in view of the effect on the jury, are the most pernicious features of the traumatic hysteroneurasthenias. It is not true, however, that these patients get well as soon as they are paid. In the long period of waiting they often develop a true psychotic state that takes many years to efface.

In the type of hysteria met with in the well-bred and intelligent, the prognosis is fairly good in the young; with advancing years it becomes less hopeful. The most favorable types are those seen in childhood.

A few cases of fatal hysteria have been reported. The exact significance of these cases is far from being settled. The longer a hysterical symptom has persisted, the more sinister the prognosis, but paraplegias, hemiplegias, of three or even five years' duration, have been known to recover.

**Diagnosis.**—If hysteria may simulate almost every known combination of symptoms, its diagnosis manifestly presents features of more than usual difficulty. Year by year, however, the analysis becomes more and more searching, and it is only with reference to certain conditions that a differential diagnosis becomes complicated.

**The Epilepsies.**—In estimating the character of the major convulsive attacks, the epilepsies come into review, although the analogies are but superficial. Much has been written of the borderland states between these two conditions, but the more they are studied, and the more known of the hysterias, for both are not entities, the less real resemblance is found. There is really little doubt that the two conditions may be present

in the same individual, and further it may occur that the same pathological process may be the determining moment for both conditions.

Sommer suggests a fourfold division of the complex cases: (1) Epilepsy, which symptomatically resembles the severe forms of hysteria; (2) hysteria, which symptomatically resembles genuine epilepsy; (3) epilepsies to which hysteria is added, as a result of the organic cause of the epilepsy; and (4) hysteria in which epilepsy occurs as a pure complication. The term *hystero-epilepsy* only renders the concept hazy, and is best avoided entirely, even in the Richer sense as a name for the major hysterics, although both Binswanger and Apelt believe in a *hystero-epilepsy*, in the sense of the co-existence of the two diseases. Even though it be known that the positive signs fail in the differential diagnosis of particularly difficult cases, yet with continued observation a diagnosis is possible in the sense of the Sommer divisions just quoted.

In *epilepsy* the signs usually relied upon to make a diagnosis in the ordinary cases are loss of consciousness, complete amnesia, injury to the tongue or body, involuntary loss of urine or feces (the former more frequent), immobility of the pupils (not invariable), clonus or Babinski phenomena at the end of the attack (also not always), exhaustion, and sleep. In the hysterical attack the loss of consciousness is nearly complete, the amnesia is island-like, or may be cleared up entirely by association experiments, or in hypnosis, the biting of the tongue or injury to the body in falling is apt to be slight, if present at all, the urine and feces are rarely discharged involuntarily, only in rare cases are the pupils immobile, and ankle clonus and the Babinski phenomena are not seen. After a hysterical convulsion the patient is rarely sleepy, even if fatigued.

**Meningitis Cerebrospinalis Acuta.**—The occurrence of lymphocytosis and bacteria in the spinal fluid and exact observation of the temperature have disposed of most of the former pseudo-meningitis attacks.

**Organic Brain Disease.**—A number of organic brain affections may be the exciting causes of the outbreak of typical hysterical attacks. Hemorrhagic meningitis, brain tumors, cerebrospinal carcinosis, septic encephalitis, syphilitic meningitis, gummata, trauma with microscopic lesions, are among the causes known to have occasioned true hysterical convulsive outbreaks. It is of moment to realize that a number of such patients with organic affections have in their early stages masqueraded as hysterical—and under the eyes of clinicians of high standing.

**Multiple Sclerosis.**—Of all organic neurological affections this offers the most difficulty in differentiation from hysteria, especially in its early stages, and the problem is further complicated since the two disorders may coexist in the same patient. The regressive character of many of the symptoms of multiple sclerosis further complicates the picture. Nystagmus, ankle clonus, Babinski's sign, and temporal disk pallor when present speak almost indubitably for multiple sclerosis. Confusion has been introduced by Van Gehuchten in his reported cases of hysteria with the Babinski phenomena. Most patients with multiple sclerosis can be markedly helped by suggestive treatment. This constitutes another occasion for question in the diagnosis. The disturbances in sensibility in the two affections may be identical: The newer studies on

the knee-jerks by Weiler have shown important differences, and the use of the association test after the methods of Jung and Ricklin throw more light on the psychical mechanisms and give further aid in the differentiation. When the two conditions seem to co-exist, the greatest care is necessary not to attach too little importance to the organic signs mentioned as being extremely rare in hysteria.

**Hemi-anesthesia.**—The differential diagnosis of organic lesion of the sensory cortex, or the posterior third of the internal capsule, is often not simple when the picture is one of a pure hemi-anesthesia. Here the search for astereognosis and for disturbances of deep sensibility must be made with great caution, avoiding the slightest intimation of suggestion in the examination. In the hemi-anesthesia of the *thalamic syndrome*, the accompanying pain, slight paresis, disturbance of deep sensibility, of postural sense, slight choreo-athetoid movements, increased reflexes, no Babinski sign, all on the same side, are the chief signs pointing to an organic lesion. Thalamic lesions, when small, have been diagnosed as hysterical hemiplegias with hemi-anesthesia, largely because of the absence of the Babinski phenomenon.

**Syringomyelia.**—Schlesinger, in his monograph on syringomyelia, has called particular attention to the vasomotor disturbances occurring in syringomyelia when limited to the posterior horn, which may resemble very closely those of hysteria. Such a localization is rare but difficulties arise in the initial stages.

**Chorea.**—Hysterical choreas offer many points of confusion. In the more frankly organic choreas, with spinal fluid findings and slight hypotonus, tendency to adiadokokinesis and altered plantar response, the diagnosis is not difficult; nor in those patients with acute infectious disease, notably streptococcus infections. There remain a number of choreiform affections that recover rapidly on suggestive treatment, but even they should not necessarily be classed as hysterical. Choreas to be considered hysterical should be accompanied by other signs of that psychoneurosis.

**Neurasthenia.**—Ziehen has given the following rather didactic, yet practically useful differentiation:

## HYSTERIA.

Mostly unilateral, patch-like or regional anesthesia, hypesthesia, and hyperesthesia, and analgesia, hypalgesia, and hyperalgesia.

Pressure points with hysterogenic and eventually also hysterophrenic characters more marked mostly in one side, usually combined with hyperesthesia.

Paralyses not unusual. Headache localized in spots, rarely a sense of pressure.

Visual fields diminished.

## NEURASTHENIA.

Sensibility intact or generally increased.

Pressure points without such characters, usually symmetrical and with intact skin sensibility.

Nearly always abnormal tire. Headache often band-like and sense of pressure frequent.

Visual fields only diminished under the influence of fatigue.

## HYSTERIA.

## NEURASTHENIA.

Smell, taste, and hearing often involved.	Smell, taste, and hearing involved symmetrically and usually in the sense of a hyperesthesia.
Skin reflexes often unequally modified.	Skin reflexes rarely different on two sides of body.
Often typical attacks.	No attacks—or rarely attack-like emotional movements set up by affect action, mostly depressive ideas.
Mood excessively variable.	Mood irritable or hypochondriacal.
Intellectual activity disturbed, especially the attention.	Intellectual activity disturbed by reason of early fatigue.
Marked suggestibility.	Suggestion influences slight.
Sleep often excellent.	Sleep mostly bad.
Course polymorphous.	Course rarely polymorphous.

**Neuralgias.**—The frequent attacks of pain of the hysterical are often confused with neuralgic pains of reflex or direct origin. The lay ideas of nerve distribution offer a clue to the diagnosis.

**Psychoses.**—Since hysteria is accompanied by, or founded upon, a modification of the entire personality, it is not surprising that hysterical psychical symptoms should be found in the non-hysterical psychoses. Nissl has stated that in from 10 to 12 per cent. of the psychoses in women this is so. It is practically true for the manic-depressive group and in the initial stages of dementia præcox, particularly those with preponderating catatonic signs. Occasionally one finds typical hysterical pictures in general paresis, and the hysteria of senile and presenile depressions is classical. The hysterical dream states are diagnosed with great difficulty from similar conditions in epilepsy, alcoholism, manic stupor, catatonia, and traumatic dream states.

So far as *manic-depressive insanity* is concerned, the accompanying signs of divertibility, flight of ideas, of psychomotor activity and the press of activity are usually sufficient to make a diagnosis. Mistakes are common, and the injudicious advice of marriage (the panacea for hysteria) has been a hideous mistake in view of the subsequent development of a marked psychosis.

**Paresis.**—Paresis should offer little difficulty. Irregular pupils, characteristic memory defect, speech disturbances, tremors, spinal puncture findings and the Wassermann reaction give definite signs.

**Dementia Præcox.**—The entire hysteria problem centres about this psychosis, and diagnostic problems arise, which at the present time are almost impossible of solution. Thus the differentiation of a catatonic stupor from a hysterical dream state may be impossible. Often the only help lies in the history. The sudden onset, after a psychical shock, speaks for a hysterical dream state, while a longer and slower course is indicative of catatonia. The foolish conduct, negativisms, catalepsy, amnesia, grimaces, analgesias, etc., are common to both conditions.

A negativism that lasts for days and weeks is evidence against hysteria, and energetic refusal of nourishment points in the same direction. Hysterics even in a dream state are less liable to soil themselves than catatonics. Unilateral pressure points and disturbances of sensibility are more indicative of hysteria, but not definitely so.

A stuporous state preceded by hypochondriacal ideas, by ideas of influence, and changes in character in the direction of emotional stupidity, or disinterestedness and menstrual disturbances, points to catatonia.

The remarkable analogy drawn by Jung<sup>1</sup> between hysteria and dementia præcox explains in large part why such a difficulty arises, and the present author<sup>2</sup> has attempted to show that the symptoms of the two conditions, although identical, are, nevertheless, representative of quite different stages in the synthesis and analysis of personality. As one in going up a mountain obtains the same view at any one spot as when coming down, so in the peculiar dissociations of personality in its still loose synthesis (hysteria) and in the dissociation of a disintegrating personality (dementia præcox) one may obtain quite similar symptom pictures. The emotional apathy of the dementia præcox patient shows a striking resemblance to the indifference of the hysterical. Jung has shown that in both there are emotional complexes, which are covered up and hidden, or repressed. In the hysterical the mood does not last long, but is suddenly interrupted by an explosion, a crying spell, muscular contortions, and the like. The impulsive acts of the apathetic dementia patient are similar, although here the protective mechanisms are harder to penetrate and the painful buried complex more difficult to reach by reason of the severe blocking.

The tendency to repress the unpleasant and to bury it as deeply as possible, which is a healthy psychological means of adjustment, is seen in its diseased phases in both disorders. In the hysterical, following Freud's interpretation, one finds the conversions, the displacements, whereas in the dementia præcox cases one finds the peculiar transpositions and blocking, which prevents adequate reactions to reality. Although Jung deprecates the use of the phrases "hysterical character" and "dementia præcox character," yet they are useful concepts, if one does not push the words too far. All types of temperament may be found among hystericals, yet it is characteristic of these personalities that a powerful emotional complex is present which is incompatible with the ego-complex. One encounters certain embellishments on the part of the dementia patients which show the influence of such emotional complexes, which are also present in hystericals. It is frequently seen in forms of studied and pretentious behavior, aristocratic gaits, philosophical enthusiasms, religious originalities, etc. These are frequent expression of either the hysterical or dementia præcox reactions. Delusions of social elevation in dementia præcox frequently manifest themselves in exaggerated manners, studied speech, bombastic expressions, affected eloquence, and high-sounding phrases. One sees in the quasi-religious schemes such as Christian Science, both hysterical and dementia præcox features.

<sup>1</sup> *Physiology of Dementia Præcox*, translated by Peterson and Brill, New York, 1909. *Nervous and Mental Disease Monograph Series*, No. 3.

<sup>2</sup> *Predementia Præcox*, *Am. Jour. Med. Sc.*, 1907, cxxxiv 157.

**Treatment.**—No pathological manifestation is in such urgent need of therapeutic individualization as hysteria, and to lay down general laws of treatment is to invite failure. The vast majority of patients suffering from the hysterical reaction can be benefited, but it requires a great nicety of adjustment of means to ends, and a refined tact and at times almost superhuman ability to choose from the various resources at hand in order to accomplish permanent results. Without a fairly complete insight into the chief etiological factors in the individual case little can be accomplished beyond the mere removal of some of the more striking symptoms.

In no nervous disorder is *prophylaxis* more important and so universally neglected. For this latter reason it should occupy the first place in therapeutic considerations. It should include a knowledge of and a contest against all the various influences that conduce to the development of the hysterical character or that encourage its further development in a susceptible individual.

The facts concerning heredity teach that syphilis, tuberculosis, and above all alcoholism form the soil in which the best hysterical products are raised. Accidents during childbirth are also fruitful sources for the development of the nervous constitution that falls a prey to the hysterical reactions. Thus, enlightened eugenics would avoid the marriage of blood relations in whom nervous affections are present, and the marriage of syphilitics and the tuberculous should be most rigidly scrutinized. Only under the best of circumstances can one minimize the real danger in the probable progeny of such marriages.

The hygiene of the pregnant mother, so essential from all other points of view, is doubly important from that of nervous inheritance. The general laws concerning hygiene of the nursing and child cannot be entered into here. Statistics would seem to show that children nursed by their own mothers run less risk of developing hysterical accidents, yet with improved pediatric methods the application of this must not be too rigidly insisted on, especially when the mother's health—not her comfort alone—suffers from the strain of continued nursing.

An unwise and unreasonable desire to stimulate the very young child is a prolific source of the nervous disposition. The nursing child should be left very much alone, and not made the plaything of every passing stranger, much less the centre of admiration of doting relatives and friends. Much has been written concerning the nourishment of the growing child, with the unfortunate result that there is a great deal of rampant faddism, and the boy or girl often gains that little knowledge which is a dangerous thing. Discussion of what the children should or should not eat, if at all necessary, should be carried on behind closed doors, and not made the tri-daily subject of admonitive conversation with the family at the table. The exaggerated fancies concerning what is digestible and what is indigestible gained by the children from such discussions at meal-time form the basis of a multiform hysterical dyspepsia in after-coming years. There is probably no single means whereby pernicious suggestions play havoc with the mental health of children, and adults as well, than the continued discussion of digestive physiology



at meals. Hot biscuits and other items of diet play a minor rôle in comparison with half-baked dietetic ideas set forth by anxious parents.

The merest common-sense notions of eating rationally and slowly are all that it is necessary for a child to know about its diet. If children do not care to eat certain things, it is extremely unwise to make a fuss about it. It only concentrates their minds on the implied importance of it. If left alone they grow out of the notions sooner than when a scene is made over every childish whim. Such commands fix the child's distastes, and prevent the free play of the important law of forgetting.

Certain articles of food should, however, be tabooed. These are alcohol and all alcoholic containing liquids. Tea and coffee are not good for growing children, and an excessive meat diet as well as an excessive fat diet is harmful. A mixed diet with well-cooked carbohydrates and abundant fresh vegetables is desirable. Avoidance of monotony in the meals will give appetite and zest to eating as well as a wider experience that tends to correct one-sided notions concerning food.

The *mode of life* of the child is in need of careful regulation, yet the machinery of such control must not always be on view. An orderly day without interruption in its specified tasks and an abundance of sleep are imperative in the ages up to about eight, or the ordinary beginning of school period. The afternoon nap for the child up to six is desirable. Naturally, children's parties, with rich food, candy, and the excitement of imitating the social whirl of their elders, are bad. Anything that overstimulates the child is disadvantageous. It has been found that single children are much more apt to be hysterical than those growing up in a large family, probably because the hotbed culture due to constant contact with older minds often develops immature fruit, especially if the younger mind is overstimulated or is spoiled by excessive indulgence. It is also a striking fact that a single brother in a large family of sisters comes out a good candidate for the hysterical reaction.

The young child of nervous organization will need special physical and mental training. Gymnastics and games in the open are advantageous; hydrotherapy is useful if not overdone. The daily cold plunge is a useful tonic, but such methods may be very readily carried to extremes. The child's enjoyment of such procedures is a better guide to their usefulness than a rigid scheme of "hardening" the children. Summer sea baths or lake baths in the open give the ideal hydrotherapy.

Play is much vaunted, but it is in need of careful scrutiny. As a rule, the childish mind needs little stimulation of the imaginative faculties. The child lives in a sort of disconnected dream most of the time, and what is needed is not forcing in this quality, but direction of it. Just how this direction shall be carried out in the individual case must be left to the tactful mother. If she be hysterical, the most difficult problems arise, for then one has both heredity and constant suggestion to battle against. Under such conditions most children do better away from home, unless the mother has gained by hard experience and careful study the lessons that help her to remedy her own anomalies of inheritance or defects of bad training.

The factor of *suggestion* is practically identical with that of imitation.

Here enter both the advantages and the dangers in the bringing up of the children. The child imitates its surroundings and adapts its imitations in accord more or less with its primary inherited likes and dislikes. These are, however, not immutable, and are made so only by the constant imitation of the parents. The children profit or suffer from the daily examples set before them, and by influencing the parents to the soundest methods of training one arrives at a scientific pedagogy that can eliminate the most pernicious hysterical weeds that a bad heredity can sow. The ability on the part of parents to be cheerful under all circumstances; to bear petty annoyances of life with equanimity; to keep their worries and angers to themselves, is a constant stimulus in the right direction for developing children.

The *school* question soon obtrudes itself, and here a host of difficulties arise, concerning which unanimity of opinion is obviously lacking. Too often one regulates the going to school by chronology, but experience teaches that no two minds are alike, and that there is a physiological age that may be different from a chronological age. In the mental sphere this is very striking, and it is sound pedagogy to attempt to teach a child this or that not according to his years or months or days, but according to his individual development. It is for this reason, if no other, that parents should know the teachers in a school, if outside schooling seems to offer the best resources, and a constant interchange of thought should take place between the school and the home.

In connection with the education of the hysterically susceptible child, particular attention should be directed to the *emotional life*. Here is the weak point in the hysterical character—the lack of emotional control; and especial emphasis should be laid upon the training of this pathological lability of the affective life. Children should not be overindulged; their minds should be directed from their little aches and pains; they must understand what danger is without fearing it. They should know what hunger and cold are, and learn to disregard both. The observance of the wishes of others, and the need for accommodating their desires to the interests of others should be tactfully taught. Simply telling children these things is not real education. The opposing affective states must be called up, and thus self-control taught. The process of physical hardening is of no value if the emotional self-control is not acquired at the same time; fortunately the necessity for action and precision required in outdoor games, or indoor athletics, is a most helpful aid in acquiring control over the activities of the sympathetic nervous system, which is the most important link in the emotional chain. A proper sympathetic nervous system hygiene does not overlook the importance of regularity in defecation and urination.

A close control of imaginative literature is essential. Highly exciting tales are to be excluded, but they are rendered obnoxious, as a rule, by an early cultivation of the young boy's or girl's ideas of style in language and good writing, as the defects soon become so obvious that such kind of writing fails to attract by reason of its vulgarity or poor style. Hysterical adults would do well to avoid reading works that stimulate day dreaming, castle building, and especially the erotic.

The young boy or girl should be encouraged to build, to dig, to carpenter, to swim, to skate, to ride horseback, and works of phantasy should be kept in the background. Much of the handiwork done by girls is bad, especially that which can be done mechanically, as it encourages day dreaming. If the stream of consciousness be directed in productive lines there can be no objection to such work. The cultivation of efficiency and productivity are in direct antithesis to the dreamy, hazy, inefficient, slipshod features of the hysterical character.<sup>1</sup>

Isolation and solitary pre-occupation are bad features for the predisposed individual, hence the question of the child's companions should be made a matter of special moment by the parents. Leaving these matters to adjust themselves by chance is usually a lazy method, yet it may be less harmful than the setting up of hypothetical, overdrawn, and impossible ideals. The real rub with companions, whereby the child must make the necessary adjustments and find itself, is more helpful if the wise parent sees the faults in adjustment and will take the trouble to work on them, than when it tries to seek an impossible degree of perfection in the environment. Young children who are wrapped in cotton wool invariably suffer from this fancied protection. Hence good public schools and the large private schools are often much preferable to the smaller schools, where, unfortunately, much snobbery is rampant and special privileges are granted. These schools may not harm the normal child, many of them are highly desirable for special training, but they should not be selected for the child with hysterical, syphilitic or alcoholic antecedents, or for those who show the development of great emotional instability. The so-called "tender" or "sensitive" child must not be handled too tenderly, nor yet be left absolutely to its own resources.

The avoidance of sexual factors even for the young child is a *sine qua non*. In practice, however, it is much more difficult than in theory. For this reason nature study has an added claim for recognition. It not only brings the young bodies into the open air, but also brings the mind into contact with real things and actual forces. Their sexual education thus becomes natural and gradual, and sexual ideas are less liable to be distorted as the period of puberty comes on.

Special difficulties arise in the training of children who show a marked tendency to morbid anxiety, fears, and self-consciousness. Those who are afraid of being left with strangers, of sleeping in the dark, of going into the woods alone, of doing anything alone for the first time, are rarely helped by harsh measures. Such methods only tend to suppress their expression and force the children into expedients of lying and even hysterical conversions. Supposed headaches or other physical ailments are complained of in order to avoid exposure to disagreeable experiences, and thus the patient is forced into a hysterical defence reaction.

The principle of prophylaxis continues just as operative in the years after puberty as before, although the personality becomes more fixed and is modified with greater difficulty, but at no time can it be said that one's

<sup>1</sup> See Payot, *Education of the Will*, Funk-Wagnall, 1909.

ideas become so firmly settled as to be incapable of some change, at least if only the correct method of approach be found.

**Etiological Therapy.**—Emphasis has been laid upon the fact that in many individuals possessed of varying degrees of the hysterical constitution physical factors may play an important part in determining a breakdown in resistance, and the development of hysterical symptoms. Many of these individuals are not really hysterical in the narrow sense of the word. They possess only that common fund of hysterical reactions with which all are born. With them, however, the stress is too much by reason of an added physical or social burden. To ascertain just what this is, and to relieve it is the problem of the attending physician. The diagnosis is often not simple, since considerable doubt may arise as to whether the physical disorder is primary or secondary.

The organs of generation in women call for special scrutiny, and although disturbances here rarely have any relation to the hysterical condition, nevertheless, in those with such a predisposition gynecological disorders may break down an acquired resistance to hysterical manifestations. Even in the average woman pelvic disease may bring about hysterical outbreaks. Under any condition grave gynecological disorders require proper treatment.

It should not be overlooked that unnecessary gynecological interference in the typical hysterical approaches the criminal, or that major gynecological operations are a frequent and direct cause of hysterical breakdowns. The gynecologist has a stony path to travel in the hysteria domain. Attention might be directed here to the clinical group that Dupré had dubbed "mythomanias," since so many of them have close hysterical relationships. Many of these patients are obsessed with the idea of operation, and invent symptoms in order to gain their point.

In hysterical virgins gynecological interference, barring definite and precise indications, is to be avoided. It almost invariably aggravates the condition, and by suggestion may become the nucleus of a "mythomantic" obsession. Too much stress cannot be laid upon this subject, for there is a great amount of harm done by gynecological tinkering.

Another consideration with even less to recommend it is that of gastric disturbance. Of all hysterical manifestations, those of the stomach are among the most common, and numerous are the attempts made to influence the mental state by way of the gastro-intestinal canal; and, strictly speaking, without results. The good results are obtained through suggestion alone, and the bad results, also due to suggestion, are legion in number. Gastrotherapy, with its faddy dietetics—speaking only from the hysteria standpoint, be it understood—has contributed more to the actual production of hystero-neurasthenic stomach disorders than almost any other form of bad medical suggestion. This may seem an extreme position, but a clarified judgment forces one into it.

One special etiological factor demands an emphatic expression. It concerns the sexual life, and more particularly *marriage*. The hoary age of that etiological factor known as the unsatisfied uterine longing, and its supposed remedy, marriage, both of which are still popular serve to show how extremely subtle the gradations are in the type of

disorder under discussion. Its justification seems to rest on the universal fact that flighty young people settle down after marriage. But there is a great difference, from the standpoint of psychiatry, between the hysterical constitution and the irregular or often so-called hysterical conduct of young people, women particularly. Marriage does not cure hysteria, and whatever judgment may be reached with reference to Freud's ideas of juvenile sexual traumata as a cause of hysteria, it is certain that marriage on general grounds is dangerous therapeutic counsel for the hysteric. The solution of the problem of a contrary position, should marriage be denied the hysteric, is also difficult. Certain writers take the position that the physician should not advise under these conditions; he should simply state the situation objectively and let the family decide for themselves. As an actual matter of fact, the specialist is called upon to advise, but his advice should be given in the form of a free discussion after all the facts of the case are available—the personality of the contracting parties, their heredity, social opportunities, and attitude toward, and capacity for, meeting their responsibilities.

**Treatment of Developed Hysteria.**—Psychotherapy expresses in a word the most profitable mode of treatment. But the word conveys but a very inadequate idea of what is meant by the general notion. It here means all those mental methods, with or without physical adjuncts, whereby the personality of the individual may be influenced to a healthier degree of self-control. Psychotherapy includes all mental influences, from the use of the cheapest charlatanism to that of the highest pragmatic philosophies. That which may be accomplished by a cheap trick for the ignorant coal heaver may have to be worked out with much labor by means of the most tactful dialectics with an educated college professor. A word, a command, may relieve a child of a hysterical paralysis, but it may require years of careful psycho-analysis to eradicate the same in a woman of intellectual and refined cultivation. The use of the command and "harsh method" for the latter would be as ridiculous as the psycho-analytical method in the former.

Psychotherapeutics is so wide that it is no wonder that it finds its practitioners in all spheres of life. Such have come into existence as a more or less direct outgrowth of the social milieu, and their teachings are more or less adapted to individual needs and ignorance. General laws are inapplicable, but there must be a fundamental principle of moral reëducation in order to obtain permanent results. One may have to run the gamut of bullying, and cajoling, of hypnotic hocus pocus, and appeal to social or religious prejudices; one must learn to play upon conceit and vanity, on love of family pride and desire for social prestige; the entire armamentarium of suggestive influences will be found necessary if one would conquer all cases, and the man is rare who can command them all. Hence in practice one falls back upon his or her natural bent, and invariably develops a one-sided or many-sided psychotherapy according to his natural endowment and acquired method.

Such positive psychotherapeutic treatment falls naturally into a few large groups, for the exclusive use of which one finds many special pleaders. The most important of these may be conveniently grouped as *hypnosis*,

*suggestion*, and *reëducation*. In using these categories it is evident that they are not to be understood as different things or as mutually exclusive, nor as exhausting the subdivisions of the subject.

**Hypnosis.**—The concept of hypnosis has changed considerably since the early days of Liebault, Charcot, and Bernheim, yet the word is still frequently understood in the sense of an appeal to the miraculous, the superhuman, or the subconscious. Under its modern definition as only a form of suggestion it loses much of its significance. As used here it is understood as a mode of impressing certain ideas on an individual's mind after having, by trick methods, induced a condition of modified consciousness, known as hypnotic sleep, the hypnoidal state, etc., not to mention the numberless variants insisted upon by various students.

First, as to its applicability in hysteria, it is generally known that some are refractory. They cannot be hypnotized; they usually represent a higher level of intelligence than those who may be hypnotized. In such its efficacy is *nil*. Among those who are most readily hypnotized one finds the weak-minded, the mental inferiors of modern German classification. The appeal to the marvellous and the apparently supernatural has a great hold upon these, but since their hysteria is due to their real constitutional mental inferiority, the hypnotic suggestions are of very little permanent value; they simply reinforce and repeat the type of suggestibility that is an essential feature of the disorder.

Another type, how numerous they are it is difficult to decide, responds to hypnotic suggestions, not on a basis of weak-mindedness, at least not in the sense of general averages, but rather as a pure expression of the hysterical personality. To obtain a foothold with these patients it may be necessary to start them with hypnosis, but with hypnosis alone one rarely cures a severe hysteria. It may be of advantage as an entering wedge, but if continued it only perpetuates the type of reaction we are trying to eradicate. Hypnosis seems to make some startling cures, but alone it does not modify; in fact, it renders the hysterical personality more susceptible. It therefore does harm as an exclusive mode of treatment. One is making real progress in the treatment of a hysteric when the patient has learned to be uninfluenced by hypnotic passes.

In the hands of the unscrupulous or the unpractised a great deal of damage can be done. The fancied influence for absolute evil that hypnotists are believed to exert on their subjects can in reality be accomplished only in the weak-minded. Such individuals do not need any pressing of hands or looking into mirrors to produce their lack of will.

It is a striking commentary on the value of hypnosis in the treatment of hysteria to find it practically rejected by its warmest advocates of twenty years ago. With the limitation thus outlined, however, it will always persist as a useful adjunct in the beginning stages of treatment for some patients.

**Suggestion.**—A definition of suggestion as used in psychotherapy is as unsatisfactory as that of hypnosis; and the difficulties have become more manifest since Babinski uttered his terse definition of hysteria as "that series of phenomena that can be produced by suggestion and cured by persuasion." In 1891 Bernheim claimed that suggestion could

do away with the hemi-anesthesia of a hemorrhage in the optic thalamus; it could cure a multiple sclerosis, or overcome the palsy of a lead neuritis. Such a suggestive faith could move mountains, but it has no such import in the present section. On the other hand, there is the idea of suggestion which says that it is "some kind of an idea that enters into the mind in some sort of a manner," as Janet puts it. Such a use reduces it to a level with credulity, or belief, or unreasoning faith. Babinski has defined his own use of the word as "suggestion should express the action by which one endeavors to make another accept or realize an idea which is manifestly unreasonable, and that persuasion ought to be applied to ideas that are reasonable or which, at least, are not in opposition to good sense." Suggestion thus becomes a one-sided mechanism; it can bring about hysterical phenomena, but can do nothing to relieve them.

For our restricted purposes of psychotherapy, suggestion consists in bringing about an affect state by influences, the import of which are not apparent to the individual. It consists in inducing mental associations leading to the modifications of the patient's emotional and, therefore, psychical state that will cause actions which make for a better adjustment. Suggestions enter consciousness, either perceived on the threshold of consciousness, or on its margin, awaken ideas, associations, and in a manner similar to an endless chain bring about what Cajal has well termed "avalanche" action, which has a compelling force on the individual, who may be, and usually is, unmindful of the origin of the influences. The wise suggestor knows how, by little hints and side remarks, by appeal to fear, to jealousy, or to praise, to cause a summation of impulses which have an impelling force far exceeding that of a command. Such suggestions may thus have a very extended action, they may govern the activity of the glands of the heart or vasomotors, they may split definite idea complexes, and may so affect the senses that illusions and positive and negative hallucinations may take place.

Suggestion used in this sense, then, is different from command or direct action, and it also differs from persuasion. A loose application of the word suggestion confuses these three procedures. Perhaps, after all, the distinctions are superfluous in practice, and the resourceful therapist makes use of all three. One may command a monobrachial hemiplegic to raise his arm; if uttered suddenly and in emphatic tones, success may crown the effort. One may urge and urge a patient day by day to walk, telling him he is getting stronger and stronger and will soon be well; this is persuasion or mediate suggestion. The hysterical bedridden paraplegic may suddenly get up and run in response to the cry of a child who is in danger; this is suggestion (indirect suggestion). The child's cry, its helplessness, the need for relief, make a continual emotional appeal of such power that the forces of inhibition are stampered, as it were, and effective action results. By casually commenting on the erect carriage, and graceful, easy walk of a fellow patient, an astasic-abasic may be immensely helped, provided the praise and commendation be wisely apportioned and carefully administered. The vanity, the desire for praise, the egotism, the ideals of a patient must be correctly estimated in order to bring about corrective suggestions.

Such suggestive treatment is usually combined with a purposeful neglect of the chief appearance in the disease, *i. e.*, so far as direct attention or questioning is concerned, and later, persuasion or complete discussion of the disorder can be added to reinforce the advance started by the suggestive ideas.

Command, suggestion, or persuasion are only symptomatic remedies; they do not change the chief factor which is responsible, namely, the hysterical character. Following the opening which they afford, a more fundamental method is necessary in order to permanently modify the personality and to make a recurrence of the manifestation less probable or impossible.

**Reëducation.**—This is the ideal psychotherapeutic goal. It aims to reconstruct the individual on a firm basis of reasonable and helpful, philosophy that permits him to understand his weakness and his strength, his limitations and his powers. Mills<sup>1</sup> has well summarized a part of its aims in saying: "This method contemplates teaching the patient what he has, what he has not, what he seems to have, what he can do, what he cannot do, and what he simply believes he cannot do." But it is more than this. It concerns itself less with the patient's illness than it does with the steady cultivation within him of that individual mental and moral stability that makes true men and women, and not molluscs.

The exposition of the aims to be accomplished needs a short summary of the methods to supplement it. In a quick review of the individual patients, one is struck at once with the fact that they are very dissimilar and require quite different treatment. One recognizes that for the imbecile and weak-minded types, for whom perhaps only a cure of symptoms can be hoped for, hypnotism may be justifiable. An appeal to the miraculous relieves for a time, but the patients usually go on their way with ever-changing symptoms. Reëducation for such is a waste of effort, of time, and may squander a pittance which might better be utilized to keep the patient isolated and away from those particular parasites who feed upon the credulity of this type. The manifold fads, cults, and quasi-religions derive their followers and endowments largely from this class.

Granted a normal mental endowment in a comparatively young individual, one has a number of expedients. If neurasthenic factors appear large in the history, an *isolation* technique is advisable. Known under various names, it matters little who first invented it, but certainly Weir Mitchell did much to give it definiteness and point, nor did he neglect the reëducation features. The patient should be isolated, separated from home and family, preferably in a suitable hospital room or ward, or private house. The choice of a suitable nurse is of paramount importance. One's chances of success, in the vast majority of cases, depend more on the coöperation of the physician and nurse than on the particular methods pursued.

The daily régime must be rigid or pliable according to the case. The majority need a rigid schedule, the very strictness is a part of their

<sup>1</sup> *Monthly Cyclopedic and Medical Bulletin*, July, 1908.



moral rehabilitation. For food one commences with milk, three or four quarts daily—keeping close watch on the weekly body weight, then rapidly rising to five, six, or more quarts, as indicated by the character of the stools and the digestion. The patient is not permitted to see anyone; she cannot write or read, but must lie in bed. The physician's visits must be frequent, and must be made an event in the day. One must bring to the sick room each and every time some new mental pabulum which can be worked over by the patient. It may be a chapter from Job, a fragment of natural history, a bit of vital human experience, a thought from Epictetus, perhaps a problem of conduct to be solved—something that remotely or directly bears on the subject in view, *i. e.*, the gaining of a new and healthy point of view of life, and the stimulation of moral fibre. The nurse throughout must have the same point of attack—not obtrusively so—and above all a cheap and boisterous optimism is to be suppressed.

Little by little, as the weight increases, reading may be introduced, but the books read need care of selection. One makes a great mistake in advising a book one does not know well. It may contain ideas that have a special affinity for the intense emotional complexes which are closely related to the patient's hysterical reactions. After four, five, or six weeks the patient may be permitted more latitude, both with reference to meals and associations with the outside world. This technique is to be found in many special manuals as worked out by hours, minutes, and seconds. To follow such too rigidly for all cases would be an absurdity. A regular schedule should be devised for each patient.

In addition to the general psychotherapeutic talks, which should not leave out of sight the emotional training as well as the more narrow intellectual, special days should be set aside for the discussion of some of the more intensely colored emotional features of the patient's history. In certain types of patients this may have to take on the character of a prolonged psycho-analysis in the sense elaborated by Breur and Freud. The tactics of these conversations require individual adjustment, and are a purely personal acquirement. Its principles may be reduced to writing, but its practice is acquired only by force of natural endowment and studied skill. The refined dialecticism of a Dubois, the charm of personality and resource of a Weir Mitchell, the hearty emotional appeal and camaraderie of a Dejerine, are individual expressions of great masters in their particular line of therapeutic resource.

The use of religious ideas and ideals in psychotherapy calls for special mention. Since religion is an expression of the emotional response of the individual to the most ancient of all suggestive factors—namely, the riddle of existence and the hereafter—it is not surprising that it should play an immense rôle in the affect of the individual. The evolution of the customs and usages of the priestly class symbolizes the expression of this emotionalism and thus sways it. Religious belief and its foundations should never be lost sight of in an enlightened psychotherapy, and the use of religious motives, of clericals themselves, is often of inestimable service. To the medical psychotherapist, however, trained as he alone is trained to properly estimate the etiological factors, physical,

mental, and moral, which cannot be understood without a firm grasp of biological medicine, it is a sad sight to see the pitiful reversion in modern times to the pre-Hippocratic priestly mode of treatment of hysteria and kindred psychoneuroses. Emmanuelism, Christian Science, Mental Healing, and the like are but faint and futile grasps at the large truths that medicine has made its own in the progress of twenty-five centuries. The successes of such practitioners are unfortunately offset by the manufactured hysterical products which such systems produce. But the practitioner of medicine is not free from responsibility in this respect.

Hydrotherapy, electrotherapy, pharmacotherapy, physiotherapy are necessary adjuncts, according to etiological requirements, but must be considered in the light of more or less temporary assistance. As exclusive mediums for suggestive influences they fall short of the ultimate aims of a complete psychotherapy; but as rational means to give physical tone, muscular activity, to correct disturbances of visceral adjustments, etc., they are necessary. The opium and hyoscyamus narcotics, analgesic antipyretics, and alcohol hypnotics are to be given with great caution, and never over any extended period of time. Iron, arsenic, and strychnine are not specifics, but are useful tonics. The salts of the alkaline earths, usually in combination with the phosphates, are indicated as quieting an excessive muscular irritability.

Valerian and specified drugs have no specific value, but it is not impossible to find combinations of the volatile oil class of drugs that have very definite tonic action on a dilapidated vegetative nervous system, and as such are symptomatically valuable. The search for remedies having definite and selective action on the sympathetic nervous centres, on the cord, or their representatives in the cortex has only just begun. The treatment of hysterical delirium may require the bromide hypnotics—it is only under rare circumstances that hyoscyne is necessary. Paraldehyde is, by reason of its extremely unpleasant taste, one of the most useful of the alcohol hypnotics.

**Analytical or Cathartic Method.**<sup>1</sup>—No therapeutic consideration of hysteria can claim completeness without definite mention of the method originally termed the cathartic method by Breur, and later amplified by Freud as the analytical method. In its rough form it may be termed the “talking it out” procedure in psychotherapy, and the usefulness of the confessional in church practice is due, in large part, to the same principles. Freud’s claim that the analytical method is one which acts most penetratingly and carries farthest is undoubtedly well justified, but unquestionably its application has its limitations. Although at first practised with partial hypnotic addenda, Freud now practically abandons such methods. He quotes Leonardo da Vinci as saying “that the art of painting consists in placing little heaps of paint on uncolored canvas where before there have been none, while sculpturing, on the other hand, takes away from the stone as much as covers the surface of the statue contained therein.” The method of suggestion acts like the former, analysis like the latter.

Freud’s technique has slowly evolved, and a presentation that is

<sup>1</sup> See Jones, Article on Psychoneuroses in *Modern Treatment of Nervous and Mental Diseases*, Ed. by White and Jelliffe, vol. i.

valid for the present may change in the near future. His early procedure was to place the patient in the recumbent position, the physician sitting behind the patient's head at the end of the lounge. The physician thus remains practically out of sight of the patient, who is then asked to give a detailed account of his troubles, and to say everything that comes to the mind irrespective of its seeming logic or sense and apart from disturbing, mortifying, or unnice suggestions. In all such histories gaps are inevitable. These the patient is urged to fill in by thinking closely of the attendant circumstances, speaking aloud all of the fitting thoughts that pass during this search ("free association"). All the thoughts are requested to be uttered, notwithstanding their disagreeable nature. The patient must exercise no critique and remain passive. It will be found that the disagreeable thoughts are pushed back with the greatest resistance. This is made all the more striking since the hysterical reaction, *i. e.*, the symptom, is the symbolic expression of the realization of a repressed wish and gives the patient some gratification. A great effort is made to retain the symptom, especially as its origin is not really perceived, and since it represents, in symbol, the individual's former conscious strivings. In psycho-analysis one attempts to overcome all of these resistances and by a series of judicious and tactful probings reconduct into the patient's consciousness the hidden thoughts which underlie these symptoms. Every symptom has some meaning; behind it there lies some associated mechanism, the origin of which the patient unconsciously or partly consciously represses. In the psychoneurotic symbol may be read the cryptic expression of the original thought driven back and hidden.

To slowly analyze and pick apart the mechanism is the object of the analytical method. One needs not only special tact for such excursions into the subtleties of the mental life of some individuals, but also a developed method of interpretation. Every act, every symbolic expression or action, lapse in speech, mannerism, needs to be carefully noted and its bearing coördinated. Freud lays particular emphasis on the analysis of dreams, since he believes that in the dream the unconscious, or the "repressed conscious," is more apt to reveal itself. Hence a careful reading of Freud's *Interpretation of Dreams*<sup>1</sup> is of the greatest value in this study, also his *Psychopathology of Every-day Life* as well as the Three Contributions to the Sexual Theory.<sup>2</sup> Hitchmann-Freud's theories of the neuroses is invaluable.<sup>3</sup> In his work on dreams he has developed to the full the chief directions along which his mind has travelled in the psycho-analytical method.

It is of the utmost importance to trace back into the earliest years the striking emotional influences that have come into experience, for, for Freud, the hysterical reaction consists in a perverted type of reaction to these experiences. As is known, the blurring, or loss of an emotional influence—an affect, in short—is due to a number of factors. In normal life forgetting is the commonest type of a corrective adaptation, and

<sup>1</sup> Macmillan Company, New York, 1913; tr. by Brill.

<sup>2</sup> Tr. by Brill, *Nervous and Mental Disease Monograph Series*, No. 7.

<sup>3</sup> Tr. by Payne, same series, No. 7.

forgetting is carried out with special ease if the emotional stress has not been excessive. Forgetting, however, is only a secondary phenomena, and usually is more successful if the immediate reaction has been an adequate one. Such immediate reactions express themselves as tears, as anger, as impulsive acts, etc., and in such reaction the affect is discharged. In every-day life one calls it giving vent to one's feelings. If, however, the reaction is suppressed, the affect becomes united to the memory of the experience, and an emotional complex, or, to use a rather broad simile, a psychic boil results, which must heal by absorption, by discharge, or by other means. Freud uses the term *ab-react* (*abreagieren*) to signify the adequate reaction, or discharge, of such affects or their resulting complexes. Talking the whole thing over, giving vent to one's secrets and confessions, are well-known *abreactions*.

In hysteria certain of these complexes remain prominent; they are neither reacted too promptly, nor is their unpleasant feeling tone diminished by the blurring process of forgetting, although it is characteristic of the Freud point of view that the actual experience which gives rise to them becomes forgotten and the cause of the affect disturbance which becomes later converted, it may be into physical signs, remains apparently unknown to the patient. It must be dug out by psycho-analysis, and when discovered catharsis takes place and the patient is cured.

It is further of importance to realize that *abreaction* is impossible for certain types of psychical shock or trauma. The inevitable, in the loss of a beloved person, is not overcome by frank discussions; social relations may make it impossible to mention the shock; or, again, it may be that it may concern itself with things which the person wished to forget, and which were intentionally inhibited and repressed from conscious memory.

In the limited space the full value of Freud's cathartic method cannot be presented. His original works must be consulted, as most of his critics apparently have not taken the trouble to read them. If truth be often stranger than fiction, certainly the charge of their being romances does not preclude their value. Experience is accumulating rapidly to show that in well conducted psycho-analysis one has the most valuable means of treating hysteria.<sup>1</sup>

<sup>1</sup> Jung, Theory of Psychoanalysis, *Psychoanalytic Review*, vol. i, No. 1. Jelliffe, Technique of Psychoanalysis, *Psychoanalytic Review*, vol. i, No. 1 et seq.

## CHAPTER XVIII.

### MIGRAINES. NEURALGIA. PROFESSIONAL SPASMS. OCCUPATION NEUROSES. TETANY.

By SMITH ELY JELLIFFE, M.D.

#### THE MIGRAINES.

UNDER this head will be discussed a series of characteristic phenomena, which are among the oldest affections known in literature, and which are here brought together arbitrarily, with the full recognition that divergent opinions are held as to their essential relationships.

#### OPHTHALMIC MIGRAINE.

**Synonyms.**—Sick headache; megrims; hemicrania; bilious headache; Hemikranie, Migräne. Latin, cephalagia biliosa; suffusio dimidians; migraine; emicrania.

**Definition.**—The difficulty of limiting this protean affection is great. It is practicable only to attempt a definition of the more classical types of the affection, neglecting numerous related forms bound up with other disorders. Taking these limitations into consideration, migraine may be defined as a periodical abnormal state in which the patient suffers from a peculiar oppressive pain in the head, unilateral or bilateral, localized or general, which develops very gradually from heaviness to dullness, to pain that is splitting, and is accompanied or more often preceded by characteristic visual signs, such as scotomata, flying specks, or partial blindness. Chilliness, depression and sensory visceral disturbances, particularly in the stomach, are also usually present. An attack may be terminated, after a few minutes, by vomiting, or it may persist hours or even days. After a variable length of time, usually following a heavy sleep, the patient regains his previous condition of well-being.

Tissot is credited with having given the first classic on migraine and his remained authoritative up to the appearance of Liveing's monograph, "On Megrim, Sick Headache, and Some Allied Disorders" (1873), although in the interim the symptomatology was becoming richer and the case analyses more exhaustive. Thus, Vater, Hennicke, and Heberden made observations upon the scotomata. Plenck, Parry, and Wollaston drew from personal experiences the picture of half-sided blindness. Schönlein and Romberg introduced the neuralgic theories, while DuBois-Reymond, influenced by the newer work of Claude Bernard, developed the hypothesis of arterial spasm which Möllendorf controverted, and postulated a sympathetic paralysis, both of which views were conciliated by Jaccoud and by Eulenberg (1867), who described angiotonic

and angioparalytic conditions. Recent monographs are by Auerbach and Jelliffe.<sup>1</sup>

**Etiology.**—A great number of hypotheses have been advanced. That which maintains that the disorder arises autochthonously, affecting that portion of the brain which is the seat of a constitutional anomaly and acts here locally has been termed by Spitzner the *central theory*. Another group of hypotheses may be termed the *centripetal theories*. These assume that some cause, toxic, reflex, etc., originates in a peripheral organ, which, in turn, acting through the circulation or the sensory nerves, or both, affects the brain cortex and gives rise to irritative processes, and this causes the attack. The *vasomotor* hypothesis assumes that the essential feature is a vasomotor spasm followed by vasomotor dilatation, thus causing anemia and hyperemia, which act on the nervous tissues and cause the headache.

The *toxic hypotheses* have many advocates, but all suggest different poisons, and these do not seem able ever to cause the condition when given under non-vital conditions. The followers of the *reflex hypotheses* successively call on the uterus, ovaries, foreskin, adherent clitoris, gastroptosis, enteroptosis, the eyes, the ears, the nose, the throat, etc. Each specialist here advocates his own particular organ as the essential factor, just as each chemist claims his own chemical, uric acid, xanthin, paraxanthin, indican, etc., to be the real cause.

The *combined* hypotheses next come into review, the toxic-reflex, the vasotoxic, the vasotoxic central theories, etc. The organs which have been declared at fault have been the vasomotor centres, the sympathetic, the gastro-intestinal canal, secretory, excretory, ductless glands, organs of generation, male and female, the various organs of sense, the eye, ear, nose, tongue, the muscles of the body and the general metabolism of the entire body, the brain in general, the cortex, the pons, the thalamus. Many authors pick out several factors; others confine themselves to one. Some faddists say it is eye strain; others ear strain; others masturbation; while others blame corsets, tight collars, or other folderol.

Spitzner<sup>2</sup> has subjected these ideas to a thorough criticism, and has advocated the hypothesis that migraine is due to a constitutional anatomical defect, namely, an absolute or relative stenosis of the foramen of Monro. This is the fundamental substratum of the migraine constitution. The occasional causing of a passive or active hyperemia of the brain leads to a hyperemia of the choroid plexus. This causes a more or less complete plugging of the foramen of Munro, with the production of an increase of pressure in one or both of the ventricles. The increased pressure on the vessels causes more distension and more pressure on the walls of the ventricles; a vicious cycle is established, and the migraine mounts to its height, until the pressure is relieved either by a spontaneous reduction or by the sudden let down in tension due to a shock reaction—such as occurs in vomiting, from the use of various vasodilators, etc.

The general hypothesis of disturbance of the vegetative nervous system accounts for most of the facts, and the position taken is that such dis-

<sup>1</sup> *Modern Treatment of Nervous and Mental Diseases*, edited by White and Jelliffe.

<sup>2</sup> *Ueber Migraine*, 1907.

turbance may be conditioned by a host of causes. The view advocated then admits that a certain amount of truth exists in many of the explanations, but maintains that a one-sided mode of interpretation is inadmissible. Unconscious psychical factors, particularly repression, probably play a very large part.

*Heredity* explains very little in the consideration of migraine. In a recent inquiry of some 3000 individuals it was found that less than 2 per cent. had never had any headaches which could not be allied to migraine. Typical attacks in any great number were lacking in perhaps 50 per cent., but close questioning revealed a few classical attacks interspersed with many abortive attacks. With such a wide prevalence then heredity is a negligible factor. It is like the doctrine of original sin.

**Relation of Epilepsy to Migraine.**—This has been widely discussed. To those who look upon a word diagnosis of many dissimilar conditions as symbols of entities, such a discussion is, perhaps, pertinent, even if on an illogical basis, but when the standpoint is maintained that there are many epilepsies and many migraines, and that both words may stand for things at times very much related and at other times not related at all, discussion is more or less fruitless. It is not impossible to understand that a profound change in the character of the cerebral circulation, which may be due to transitory causes, and which may give rise to an attack of migraine, may also, in rare instances, be an exciting cause of a motor disturbance sufficient to set free an epileptiform convulsion. In such a sense the subject may be discussed. An increasingly greater number of atypical and borderland conditions are coming into review, and in the more recent analyses convulsive attacks indistinguishable from the hysterical and schizophrenic motor reactions are being described. Until a clearer conception is gained of the mutual relations of cerebral circulation and motor discharge it is folly to attempt to review the interesting, though fallacious, suggestions of Féré, Möbius, and others, who claim a necessary relation between migraine and epilepsy. From the standpoint of organic brain disease as a factor in the causation of a migraine the discussion is of much service. From the psychogenetic viewpoint there are close relations between some migraines and some epilepsies.

**Abortive Attacks.**—It is difficult to be certain of the character of many conditions in patients who suffer from migraine, as there exists an involuntary tendency to drag every symptom within the net of this one great malady. Experience shows, however, that sufferers from migraine possesses a keen insight into their undeveloped or abortive attacks, and after years of observation are able to recognize them with precision. Incomplete or abortive attacks may be said to be the rule rather than the exception, and all attempts to classify the disorder according to the number of symptoms present lead to confusion.

The commonest abortive attacks are those that begin in the classical manner, with chilliness, perhaps with pinched face and cold extremities. The patient then has the scotomata and wretchedness, depression and apprehension, and then while waiting for the headache the patient notices that it does not come, and, although many may still have heaviness and discomfort, the feeling of relief is sufficient to make them feel well.

Others have added the sensation of pricking in the fingers, numbness in the hand or arm, or other sensory disturbances without the headaches. In some the entire attack will consist of a disturbed painful sense of discomfort, without sensory symptoms, scotomata, or headache, but they feel sick at the stomach, and have an attack of what they term "biliousness," which clears up after vomiting. This feeling will recur with sufficient frequency, and at times be combined with such other symptoms of a migraine attack, in its varying aspects, as to stamp the whole process as a variant of a true attack. Isolated attacks of vomiting, as the sole expression of a migraine, are known.

Attacks of scotomata occur alone, without antecedent distress, and no after-effects are noted. These are most uncommon, and historically it may be noted that Parry and Airy had such attacks. It is highly probable that the majority of patients who have had many migraine attacks will have had some of this nature. Attacks of scotomata and vomiting occur without headache. In many, headache is the only symptom.

Some patients have attacks of hemiparesthesia, without any other symptoms of migraine. Such generally occur at night, and usually follow severe mental exertion, in one patient under observation a severe ordeal in playing a difficult piece of music will bring on such an attack without other signs. This patient's severe attacks are very extreme.

Under the heading of equivalents, Liveing speaks of stomach attacks associated with some of the vascular phenomena of migraine; glossal spasms are also mentioned by him. Attacks of giddiness, vertigo, intestinal colic, mental anxiety and depression which occur at periodic times in partial association with migraine symptoms, are also noted as equivalents. There is need of further study of these isolated phenomena associated with disturbances of the autonomic nervous system.

Attempts have been made to determine the relative frequency of migraine attacks with and without the *visual signs*. These are not over reliable, because of the vast preponderance of abortive attacks over those of the complete classical type. It may be recalled that many authors have attempted to create a special type of eye migraines which are autonomous, but the gradations are so numerous when different individuals and differing attacks in the same individual are considered that such a view receives little support.

Möbius expresses the opinion that the percentage of visual accompaniments with the attacks is usually overstated. His statistics show 130 cases, with 14 visual aura. In Liveing's 60 patients, 37 suffered from scotomata. Gowers said that the cases are about half and half, with and without eye signs. It is difficult to state an individual position, the results of personal inquiries having been so diverse. Close questioning has revealed the fact that at some time or other in the course the majority have had visual symptoms, and it is not improbable that the usual statistics are largely derived from studies of too few attacks, *i. e.*, largely from the severer attacks only. Some notes on individual histories are of interest. Several patients have kept fairly accurate records of their migraine attacks for several years. One shows 168 attacks in a period of about ten years; of these, about 100 were abortive attacks, the vast



majority of which, 60 per cent., consisted of scotomata alone. Of the 68 remaining attacks, about 50 per cent. were ordinary hemicrania, lateral or bilateral, without scotomata, the others ophthalmic migraine, usually unilateral and with scotomata. Not one of the attacks was ever accompanied by vomiting. Two were associated with aphasia, fifteen with sensory tactile associations; there were five or six attacks of hemiparesthesia, one in the day-time, the rest at night. Spasms of the orbicularis were a common accompaniment.

Similar results in about a dozen other cases show records of from 50 to 200 attacks. It is rare that a patient suffering from migraine of any severity goes through life without some one or more of the sensory phenomena, and notably the scotomata. But a great many individuals have their attacks, both mild and severe, while asleep, and may never know of the occurrence of scotomata. Many of these patients, who have given no history of scotomata, have had frequent night attacks with sore eyeballs and sore spots on the scalp, two symptoms of frequent, although not necessary, relationship with other sensory symptoms.

**Classical Migraine.—Early Symptoms.**—These may be termed precursors to a full attack of migraine, or they may constitute the symptoms of an abortive attack. The most striking are a sense of heaviness, with yawning, chilliness, dizziness, or depression, motor twitching, even sharp spasms, closure of the eyelids, sensory phenomena, chiefly paresthesia, occasionally anesthesia, and affections of the eyes or other sensory organs, ringing in the ears, blowing, modifications of taste, smell, touch, etc. The temporal arteries are often smaller, and the saliva diminished.

The premonitory signs, which show a great degree of variability in different individuals, and also in different attacks in the same individual, may be felt several minutes before the attack, in some rare instances even hours. This is frequently the case in women in whom the onset of the menstrual function seems to bear some relation to the attack. At times such attacks of depression and anxiety, combined with a sense of chilliness and dizziness in the head, will constitute the entire picture of an abortive attack. Many attacks come unheralded. Many patients having attacks at night find themselves tired, and heavy, with sore spots on the scalp in the morning.

In all probability premonitory symptoms of some type are invariably present; when thought to be absent it is because the patient has overlooked them, either by reason of the mild character, or because of naturally poor powers of observation. Many patients, who have had migraine for years, have never noticed their one-sided localization, or the well-known fortification spectra, until their attention has been directed specifically to them. Many patients will deny ever having had zig-zags of light, etc., until shown Airy's pictures, when they remember having seen such phenomena. It is because of such poor observation that many cases are overlooked, which fact lends further support to the personal belief that this disorder is much more prevalent than is usually supposed.

**Sensory Symptoms.**—In the more classical attacks the patients have preliminary sensory symptoms. These are spoken of by Möbius in the sense of an aura, but the term is objectionable—even misleading. A

sense of coldness and chilliness is one of the commonest sensations. This is usually general, and is associated with a pale countenance, goose flesh, perhaps clammy hands, and a sense of misery. Cases are known, and are by no means uncommon, in which the chilliness has been one-sided, and is accompanied by other phenomena involving the entire half of the body of the same side. Yawning is a common early sign.

Unilateral *paresthesia* is not an uncommon early sensory sign. Many patients note a tingling or numbness in the fingers of one hand; this may spread up the arm, and in rare instances general unilateral paresthesia of a very uncomfortable nature may be present. In some instances such unilateral paresthesia has constituted the sole symptoms of an attack, save for the heaviness and usual discomfort. Occurring at night, such attacks are often extremely wearing, keeping the patient awake. Photophobia, flow of tears, strange sounds, tinnitus, peculiar odors, queer tastes, peppery or flat, may be noted.

*Anesthesia* is less often observed, largely because of the negative character of the symptom. When involving the face or mouth it is complained of. Anesthesia frequently follows the tingling of the early paresthetic disturbances. Franz<sup>1</sup> has shown that there is a very evident decrease in the pain threshold, especially after the headache has set in.

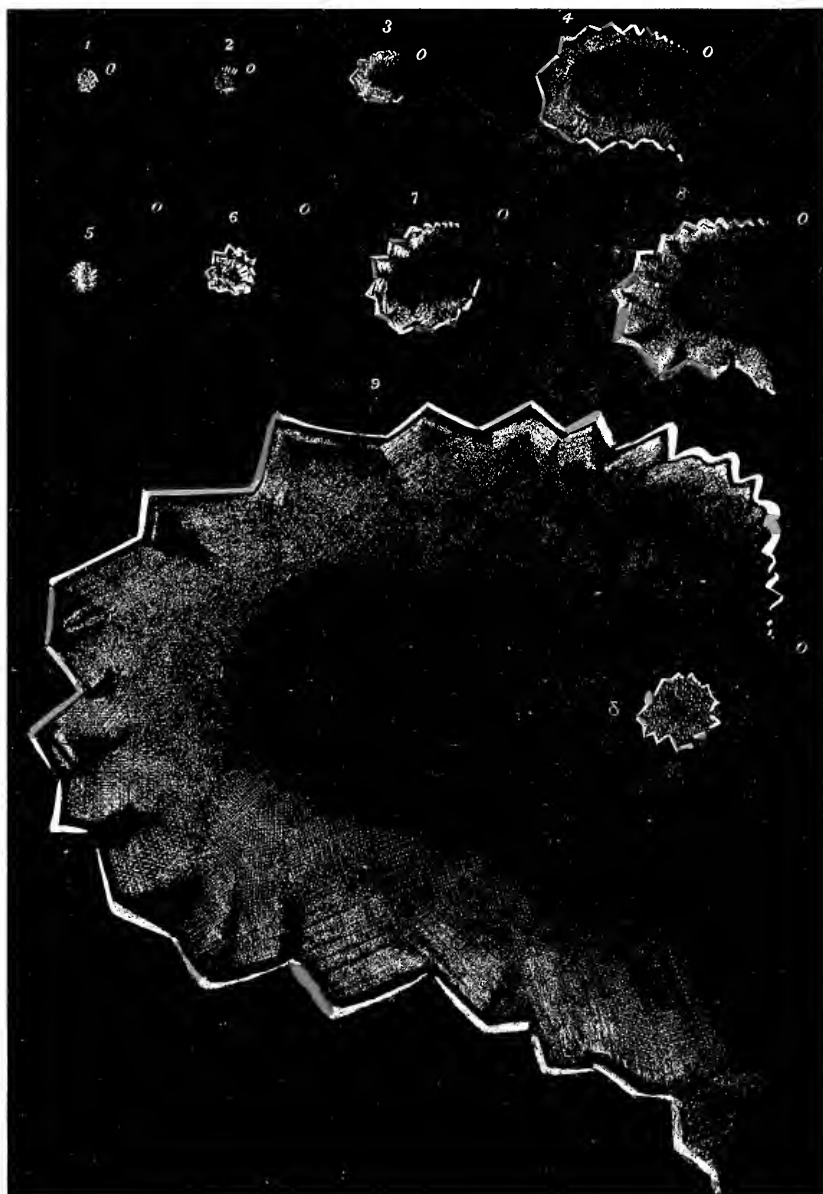
The *visual phenomena* are the most striking, and hence held to be of the most frequent occurrence. The ease of observation in part accounts for the usually accepted opinion that they are the commonest of the early symptoms. Very few individuals have been subjected to a careful sensory analysis. If more were investigated, it is probable that other slight sensory signs would be found to be equally prevalent and as evanescent. The visual signs have been described by many writers, and many illustrations have been made showing their chief characteristics. The extreme uniformity of their general character is striking, as well as the variations of the same pattern.

As a rule, the patient notices a slight blurring of his vision if reading, or a slight flicker of light located in one eye, to one side of the centre. Closer observation reveals either a slight cloudy spot, which seems to follow the eye in reading, cutting out the after-images a letter or so from the point of the centre of vision. The slight subjective sense of difficulty in reading may precede the discovery of a scintillating spot, which becomes visible on closing the eyes. Little by little this spot spreads out, usually in a crescent-like fashion. General statistics are thus far unavailable, but personal experience has shown that the majority of these scotomata have begun in the left eye, are situated to the left of the middle line, with the convexity of the crescentic border to the left. As the crescent gradually grows larger, the difficulty in seeing clearly becomes more marked, especially on the periphery of the visual field. For most, the scotomata is in constant motion, flashing in its spectral zig-zag fashion, thus giving origin to the classical name "fortification spectrum," from the play of colors, and the fortress-like "ins and outs" of the outline.

After a variable time, from five to twenty minutes, the scotomata

<sup>1</sup> *Am. Jour. of Physiology*, 1905.

# PLATE X



From Dr. Hubert Airy's Paper, On a Distinct Form of Transient Hemiopsia, Philosophical Transactions, 1870, p. 247.

Figs. 1 to 4.—Early stages of Fortification Spectra as seen in dark. O=sight point.  
Figs. 5 to 8.—Similar series, beginning lower.  
Fig. 9.—Fully developed.  $\circ$ =secondary attack within.



gradually subside, or suddenly disappear, to be followed by the headache. Not infrequently the headache never comes, and the preliminary sensory phenomena of chilliness, heaviness, and scotomata constitute an abortive attack. The classic of Liveing reproduces the excellent illustration of Airy's which is here shown.

Occasionally the right half of the field is involved. Sometimes it is the upper half, one of Möbius' patients saying that everybody seemed headless; occasionally, it is the lower. In rare instances the patient complains of total blindness, *i. e.*, central scotomata. Berbez reports an interesting case of ring-like scotomata—the patient, on looking at his watch, could see only the central pin where the hands were united; the figures on the dial were all obscured by the scintillating scotomata.

These scotomata are usually bilateral phenomena, they may begin in one eye before appearing in the other, and be somewhat different in the two eyes, and may disappear in one eye sooner than the other. Scotomata limited to one eye are probably rare.

The retinal changes during the time of the occurrence of these scotomata are uncertain. Blanching of the papillæ has been observed by some (Galezowski); pulsation of the retinal arteries with dilatation by others. Personal experience has shown similar dilatation in a few cases, but, as a rule, a normal fundus is found. The picture seen will depend upon the stage of the attack and upon its severity.

Pupillary dilatation occasionally occurs. Slight irregularity of the pupils during a severe attack of an ophthalmic migraine, dilatation being usual on the affected side, is not unusual. Bilateral pupillary contraction is the rule in the acute stage of the headache.

During the onset of the fortification spectra, it not infrequently happens that mild motor phenomena occur in the eyelid of the side to be affected. The eyelid droops a particle, and Gowers and others report double vision, interpretable as a sign of paresis in an ocular muscle. As these phenomena are not rare in the severe types of migraine, called by some ophthalmoplegic migraine, their consideration will be deferred.

**Motor Disturbances.**—*Speech.*—This may be considered as both a motor and sensory phenomena, for the most frequent type of change is sensory aphasia in most instances. Anarthrias are known, especially in the ophthalmoplegic variety, but for ophthalmic migraine the type of aphasia observed is very characteristic. As described by Charcot, it is an intermittent, halting aphasia. At one moment the patient can get the right word, at the next he cannot. He stumbles on a word; uses *madame* for *monsieur*, etc. In Liveing's cases, 15 out of 60 had speech disturbances; one, on hearing clock bells, was unable to inquire what they were. Féré cites the case of a coachman who forgot where he was going to drive his passengers; Berbez, a like case in which a pedestrian lost his way, as he could not read the street signs understandingly. Gowers speaks of a case of word deafness. Cases of agraphia are also known. Möbius reports a case with typical scintillating scotomata at one time on the right side, and at another on the left. When the patient suffered from a right-sided scotomata he had sensory aphasic signs, but they were not present when the scotomata was on the left side. The disturbance sometimes

resembles a paraphasia, the patient using a jumble of words. In a personal observation the patient could not sing a well-known tune correctly, his sense of musical values having been interfered with.

The *onset* of the aphasic disturbance may vary greatly. It is usually temporary, persisting at times not over a few minutes, again persisting a few hours. It frequently antedates the headache, or is coincident with it. In a reported case of Meige the aphasia persisted as long as the headache, and disappeared, as a rule, when that disappeared. The patient showed a loss of ability to say certain words and a tendency to the employment of incorrect words. There was no anarthria. The symptoms in one of Meige's patients were closely similar to those of a case reported by the writer.<sup>1</sup>

*Paralytic Phenomena.*—Attention has already been called to the rare occurrence of hemiparesis, which may even involve the facial muscles. Up to the present time no instances of crossed hemiplegic type have been found in the literature. This of interest in connection with the hypothesis of the bulbar origin of migraine, especially of the ophthalmoplegic variety.

*Cerebellar Symptoms.*—Oppenheim has called attention to a cerebellar hemicrania in a patient in whom every attack of migraine was accompanied by typical cerebellar symptoms. The patient was uncertain in his gait, walked like a drunken man, was dizzy, and had the sensation that the body or individual parts of it were doubled. The sense of equilibrium was disturbed in each attack. Dizziness and loss of the sense of equilibrium are not infrequent, but such a complete syndrome has been described only by Oppenheim.

*Headache.*—This is the most common feature, and exhibits a great amount of variability as to location, quality, intensity, and duration. In the more classical attacks the headache begins on the average about fifteen to thirty minutes after the appearance of the scotomata or other sensory phenomena. It frequently begins on one side, and may remain so or become bilateral. As a rule, it is frontal, or occupies the vertex, but may involve the temporal regions or the occiput, sometimes as low down as the neck. Gowers' experience points to the parietal region as being oftenest affected, and usually over a small area. Henschen, in 123 patients, shows the pain to have located 110 times in the forehead, 100 times in the parietal region, and 54 times in the occiput. There is usually pain over the eyes, and the eyeballs are usually painful to pressure. In a few instances pressure over the malar bones is painful, and occasionally there is a well-marked jaw ache.

Statistics of the percentage of location are difficult to present, since the individual will have all the different varieties. Thus, in a case already cited, in which the abortive attacks were so frequent, the headaches comparatively rare, the strictly unilateral headaches were only 5 per cent. of the entire number. In others the hemicranic type runs much higher. In Henschen's records of 123 cases, 56 had one-sided attacks, in 67 both sides were involved. In Living's 61 patients, 17 had one-sided attacks, in 7 the attacks were variable, while in 34 both sides were involved.

<sup>1</sup> *New York Med. Jour.*, 1906, lxxxiii, 33.

Möbius and others note that the headache often begins on the side opposite to the beginning of the sensory aura. Personal studies do not confirm Möbius' statement. Averages one way or another do not occur if a summary of all attacks is attempted instead of picking out those which are most vividly present in the memory of the patient. It does seem, however, as first noted by Liveing, that one-sided sensory symptoms are oftener accompanied by one-sided than by bilateral headaches. With bilateral sensory phenomena, scotomata, etc., bilateral pains are the commonest. In many attacks the pains are limited to the eyes, the soreness of the eyeballs being so marked that it is painful to move them. Pain in the neck may also cause the desire to hold the neck rigid.

The *character* of the pain defies analysis, since descriptive phrases are used in such various ways by different observers. In some attacks the head simply feels slightly sore, or heavy, or dull, or thick; "like a block of wood," is a frequent expression. "Filled with sawdust," one patient says. Again, the pain is agonizing, impossible to describe. Some patients shriek with the pain, become hysterical, and roll about the floor, grasping the head between the hands, wishing to beat their brains out. Between these extremes numberless gradients are found among different individuals, and in different attacks in the same individual. Nearly all patients will say that the severe pains are throbbing or thumping, usually indicating great pressure from within or without; as Möbius has said, "some patients think the head will burst, others that it is being squeezed in a vise." Descriptions of bursting are more common. The pain is an all-prevailing one, gradually mounting to a maximum, then running along continuously without any break, with, at all times, sudden accessions, especially on movement, if one leans over, or is forced to sudden exertion. In but the rarest instances is it described as lancinating in quality. It is the type of pain apparently seen in cerebral tumor, in acute hydrocephalus, in cerebrospinal meningitis, and allied to the pain of opium poisoning, or of sea-sick headache; all pointing in the direction of a modification of intracerebral pressure, at times an increase, or it may be a decrease, either of which may cause severe pain. Occasionally the phenomena of a bilateral headache with marked predominance of one-sided pain will be observed.

The *severity* of the pain may be conditioned by a number of factors. Movement uniformly increases it. Bending over becomes impossible. The first movement on lying down is usually accompanied by a sudden rise in severity, but this gradually subsides. The taking of alcohol usually increases the severity of the pain, as does also the use of tobacco. Eating, if possible, may help somewhat, but usually augments the pain, and is avoided. Strong sensory impressions invariably increase the pain. Noises of various kinds often aggravate the pain tremendously and cause certain patients marked distress. Möbius notes that the rage of migrainous parents directed toward their noisy children often resembles a pathological hatred. Strong light is invariably avoided, because of its tendency to increase the pain. The movements of the eyeball and attempts at visual accommodation cause an increase in the pain.

Psychical effort is often impossible; in milder attacks the awakening

of a strong mental stimulus may make one forget the pain. Möbius says that his attacks, usually light ones, are frequently forgotten during an interesting visit to the hospital, to be once more prominent afterward. The writer has frequently begun a lecture with a severe migraine to find it almost forgotten until the close, when it reappeared, usually with renewed vigor.

The movements of straining at stool, and vomiting, coughing, etc., invariably cause a rapid and sharp rise in the severity of the pain. Sensory stimuli may have an unpleasant effect. Thus, certain odors cause distress; the smell of cooking accelerates vomiting. Certain skin phenomena, such as painful spots, are frequent after the headaches.

In certain personal experiments with drugs the following have invariably increased the headache within a few moments: A few whiffs of chloroform or of ether, adrenalin by mouth, digitalis, strophanthin, and ergot. Drugs that raise the blood pressure, in general, increase the pain especially when taken at the beginning of the headache.

The headache may clear away very suddenly after an attack of vomiting, or it may pass without vomiting; in some it fades away gradually. It may last a few minutes, a few hours, or a few days. Some cases of what Möbius chooses to call *status hemicranicus* are recorded.

**Vasomotor Disturbances.**—Practically all attacks of migraine are accompanied by visible vasomotor disturbances. The battle of the adherents of the vasoconstrictor *vs.* the vasodilator hypothesis lends point to this observation. In most cases vasoconstrictor phenomena (coldness, paleness, goose-flesh, etc.) precede, to be followed later by vasodilator changes. Thomas contributes a statistical study of 107 cases in support of the early pallor, small pulse, and coldness, which pass over into the phenomena of warmth, red, flushed face and skin, and full pulse. The period of initial constriction may be unnoticed by reason of its transitory character. In some instances this initial vasoconstriction may be very marked and give rise to the phenomena of localized cyanosis, even advancing to the picture of the constriction phase of the Raynaud disease type.

In the same manner the secondary vasomotor dilatation may pass the bounds ordinarily observed and lead to localized oedema, to the erythromelalgia type, or, exceptionally, to hemorrhagic phenomena in the conjunctiva, ocular tissues, or even in the walls of the stomach.

**Secretions.**—Alteration in secretory functions are frequently observed early or late in the attack. Reference has been made to the excess of secretion of tears as a frequent precursor. Vomiting of frothy mucus, liquid diarrhoea, increase of sweat, coryza (Calmeil), or incessant salivation (Liveing, Tissot) are common phenomena. The changes in urinary secretion have attracted careful attention and given rise to many hypotheses. The early vasoconstriction of the periphery, coldness, lack of secretion, of perspiration, etc., account in a purely mechanical way for the increase of urinary secretion in the early stages, and also for most of the phenomena interpreted so one-sidedly by Haig and others.

**Trophic Disturbances.**—These have been reported by several observers. Cornu says that nearly all of his cases of migraine show facial asymmetry, and facial atrophy is recorded. These instances are largely coincidences,



and are not necessary attributes of the migraine. A facial atrophy which could be interpretable only on the basis of a migrainous disturbance of the vasomotor apparatus is very problematical, and certainly Cornu's results are not confirmed by others. Loss of weight in the severe rapidly recurrent cases is due to disturbances in general nutrition due to gastric, rather than to other causes. Herpes zoster is a not infrequent accompaniment of some cases, but the recognition of its infectious nature successfully disposes of its essential relationship to migraine.

**Psychical Disturbances.**—These have been noted by many observers, Liveing being one of the first to point out the relationship of disturbed psychical states to the attacks of migraine. Möbius takes the position that such psychical disturbances are largely due to the pain, and are epiphenomena, as it were, in many instances being related to hysteria. This position is not tenable in view of the many observations pointing in other directions.

In the vast majority of migraine attacks there are no mental changes, either before, during, or after the attacks. Mild depression, hopelessness, despondency with clear consciousness, are frequent mental states. With very severe pains Möbius admits the clouding of consciousness, and is not sure that the severe stuporous states are not due to pain as well. Mingazzini, on the other hand, believes there is justification in erecting a special group, which he has termed the hemicranic dysphrenias (*disfrenie emicraniche*), and he distinguishes a transitory and a more permanent variety. Recent observers are practically in accord in showing that severe mental disturbances varying in character and intensity may be part of a migraine attack.

Guidi has amplified these observations by reporting the history of a number of cases in which the patients suffered in the day or hours before the onset, in a much more decided manner than the feelings of anxiety or of depression described by Liveing. Thus Guidi calls attention to grave alterations in the psychical state of a number of his patients. In one the entire character of the personality would change preceding the attack. A patient who had always been calm, reserved, quiet, and modest, suddenly became much agitated, was forward, noisy, and loquacious, and told salacious stories, which was far from his usual behavior. While in health a spare eater, preceding an attack he suddenly became very hungry, and hankered especially for starchy foods. During the attack the patient had glycosuria, which disappeared later.

With the onset of pain the picture is less clear, yet there is little doubt that many patients suffer from profound psychical disturbances, which arise independently of the pain. One such case which is typical, under personal observation, would be interpreted by Möbius, and rightly so, as one in which the pain is the first link in a hysterical reaction. But there are other cases which do not belong to this group. Mingazzini's hemicranic dysphrenias may be cited as examples, in part, at least. In others severe disturbances have occurred, such as states of anxiety, rising to actual anguish (Charcot); phobias of inability to perform acts (Cornu-Charcot); terror (Liveing, Féré, Kraft-Ebing); hallucinations of sight (phosphenes, colored lights, animals) and hearing, with mental

confusion (Forli, Mingazzini); maniacal excitement (Mingazzini, Jelliffe); and stupor, unconsciousness (many authors). Liveing reports that 25 per cent. of his cases showed psychical symptoms. The Italian observers record fewer, but it appears that at least from 10 to 15 per cent. of the cases of grave hemiplegia show some distinct mental disturbance in some one or more of their attacks which is more significant than the usual universal depression.

**Symptomatic Migraines.**—The occurrence of migraine-like attacks accompanying, or due to, definite disease condition, notably organic disease of the brain, is well known. The association of migraine with gout and malarial affections has been noted. So far as *gout* as an etiological factor is concerned, Möbius is inclined to see nothing more than a coincidence; while, as for *malaria*, he holds it to cause an orbital neuralgia, not a migraine. As for the latter, it seems clear that the well-known effects of malarial infection on bloodvessel tonus are entirely sufficient to cause a typical migraine attack. It is known that attacks of migraine may be very frequent during the continuance of a malarial infection.

Migraine-like attacks are not infrequent in *cerebral tumor*; they may appear periodically, as in cases fully reported by Abercrombie and Möbius, or they may be continuous and distinguishable with great difficulty from the pain of tumor, as in cases reported by Wernicke, who has said that such attacks may be quite readily confused with those more typical of tumor. In tumors, however, vomiting brings little or no relief; quiet gives less relief, and the fluctuation in the intensity of pain is less prominent. A primary onset of migraine-like attacks in adult life should always awaken the suspicion of an organic brain lesion.

Oppenheim has called particular attention to the occurrence of migraine-like attacks in the onset of *tubes*; Möbius is inclined to think it a rare combination, and regards such either as a pure coincidence or a migraine-like neuralgia. In *general paresis*, migraine-like attacks may be an initial symptom.

**Diagnosis.**—The difficulties appear in the consideration of ordinary headaches and in neurasthenic headaches; in distinguishing between the scotomata of migraine and other scotomata; the paresthesia of migraine and other paresthesias; the aphasia, the vomiting, etc., as seen in migraine, and the same as due to other causes. In most individuals abortive and incomplete attacks are the rule, and it is often extremely difficult to determine the precise significance of these attacks.

Möbius has suggested that the problem is not only whether the case is one of migraine or not, but whether it is migraine alone, and not something additional. This author's contention that migraine is hereditary and begins in youth, would seem to make it a simple matter, but clinical experience shows that real migraines do appear in later years, apart from other affections, and as for the heredity factor, the extreme prevalence of the affection makes it difficult to accurately weigh this factor. If to heredity we strictly apply the Mendelian criterion, that the absence of the disorder in either progenitor negatives its appearance in the descendant, then the Möbius doctrine falls; if, however, by going into the

collaterals, as is so frequently done, then almost every disease having a large incidence may be proved to be hereditary.

The periodic recurrence is a difficult criterion. There is usually no difficulty in diagnosing the classical attacks from simple headache, but at times such a differentiation is impossible. Many chronic sufferers from migraine know well their real attacks, are able to distinguish abortive attacks, and also have headache of an entirely different nature. The simplest test separating abortive migraines from simple headache is the occurrence of sensory phenomena, other than pain, which have their main seat in vasomotor disturbances. It is on this account we would ally the severe headaches following the use of alcohol, ether, chloroform, opium, or related drugs to the migraines, rather than to simple headaches. The headaches of neurasthenia, anemia, syphilis, lead poisoning, nasal sinus involvement, supra-orbital neuralgia, nephritis, eye strain, glaucoma, etc., should present little difficulty.

**Treatment.**—The treatment of the migraine attack is, for the most part, fairly satisfactory. There are few patients for whom some relief cannot be obtained, both with reference to the diminution in the number of attacks, and to the mitigation of the severity of the attacks themselves. The migraine habit, constitution, or liability—call it what one will—exists in very varying degree; in some a very slight disturbance is sufficient, to set free those forces which culminate in an attack; for others it requires a very much greater mal-adjustment. If the general reflex vascular hypothesis be taken as a tentative explanation, it is very readily understood why the taking away of various forms of peripheral irritation may result in eliminating one or more, and in certain instances all, of the causes which set the migraine reaction in operation.

It is folly to shut one's eyes to the very evident clinical fact that certain migraines are relieved, if not entirely wiped away, by the correction of some peripheral disorder, sometimes more than one, which has had definite action on the nervous system. Just what the differential relation may have been between the severity of the irritant and the mildness of the attack is impossible to judge, but certainly the relief from eye-strain, from diseased turbinates, from adenoids, from constipation, from dysmenorrhœa, from a number of minor yet definite peripheral irritations, will relieve a certain number of patients. One should, therefore, eliminate at the outset such of these structural defects as are shown to have some influence on the nervous system. In denying any possibility to these influences in the causation of migraine one errs almost as badly as when maintaining some one of them to be the only element.

Gastro-intestinal factors are closely analogous to those just mentioned. In the minds of most clinicians, and certainly as generalized in the feelings of most of those afflicted, it is in the stomach, liver, or intestines that the main seat of the trouble is to be sought. The gastro-intestinal factor is undoubted in many cases; it may be exclusively gastric or colonic. As to the significance of chemical features, resulting from altered gastric secretions or from toxic bacterial products, we are entirely in the dark. It is certain that none of the products which have been held responsible as auto-intoxicants are universal causes. At any rate, the general features

of gastro-intestinal hygiene should be carried out. Constipation is to be avoided and such diet taken as experience has shown is individually applicable. Excesses in certain articles of diet are held by many as exciting causes; in many, such empirical feelings should be respected; the patient often knows himself better than does the physician.

In some, excessive carbohydrate intake acts disastrously; in others, wine, whisky, or gin. The history of inability to eat fatty food, particularly sausages, is not infrequent. In rarer instances, one notes that certain auditory stimuli may bring on a migraine. To attend certain fatiguing and thrilling operas is followed in some by migraine attacks.

For attacks conditioned by outside factors, such as malaria, gout, lead poisoning, etc., it is evident that these should receive attention.

If the varying elements mentioned have any real relation, it is evident why such a variety of measures will be of help to a few, and why so many more will be worthless for many but useful for some. Medication between attacks is largely useless, save naturally in the symptomatic migraines. General medication, for no definite purpose but just in hope that it may do some good, as iodides, bromides, strychnine, etc., is senseless. If definite factors are found that need correction, and can be so modified by drugs in the desired direction, then they will prove useful. Thus iodides will undoubtedly help many presenile arteriosclerotic migraines; bromides are useful for sleepless and irritable conditions which provide a good foundation for the nervous instability; laxatives are called for if persistent constipation bears any causal relationship.

Complicated systems of diet have been devised. Usually such are more prolific in engendering semi-invalidism than useful for migraine. Here and there a patient derives benefit from a strict dietary régime, but unless there are real reasons why a patient should not eat red meat, or tomatoes, or other sundry articles, as determined by actual experience and under repeated experimental trials, in order to eliminate faddist's errors, the patient is better off without a diet card. The reasons sought for are not those contained in many treatises on dietetics, in which medieval notions concerning differences in red meat and white meat, vegetables growing under the ground and those above the ground, are foolishly perpetuated. The only satisfactory manner to attack the metabolic problem is to carry out a complete metabolism analysis.

The avoidance of alcohol and tobacco, while advisable, is so only relatively. The individual's reaction to all influences should be rigidly estimated before those usually self-evident restrictions are imposed in the name of health.

Emotional factors are at times the most important. Very frequently migraine is a pure psychoneurosis and needs psycho-analysis.

For the treatment of the *attack* itself one finds that a like fitting of remedies to the individual is called for. In the initial phase of vasoconstriction a number of vasodilators are of service, although their action is extremely unequal. The nitrites and nitrates have been employed for years, and usually with a fair degree of success if the dosage and individual member of the group be correctly chosen with reference to the severity of the attack. A mixture is of greatest value; nitroglycerin

and erythrol tetranitrate give the best combination, for following the very evanescent and powerful action of the former, the more prolonged and steady action of the latter maintains the effect. The slowly acting nitrites are practically useless. Nature's readjustment, vasodilatation by vomiting, etc., has already reduced the cerebral pressure, and the stage has passed when the dilating remedies might be useful. It is practically only in the vasoconstriction stage that the nitrites are worth much; and in many they are inefficient. Given too late, they overdo the dilatation and increase the difficulty.

The analgesic vasodilators have come to occupy the front rank. The precise significance of each must be appreciated in order to obtain the best results. Solubility, time of absorption, slight differences in the chemical formula and in action, continuance of effect with minimum by-effects, are all to be studied. The list is a long one and is constantly on the increase. Antipyrine, acetanilide, phenacetin, and the related salicylic acid (aspirin, etc.) compounds are the chief members. It is to be remembered that while their general action is closely related, there are specific differences in the working of each, and the measure of success that one has in mastering the majority of migraines depends upon a knowledge of these factors. Antipyrine, by reason of its rapid solubility and quick action, occupies an important place, but is not always applicable. Acetanilide, alone or in combination with other analgesics of related type (salicylic acid derivatives), bromides, and caffeine are also valuable. The dosage should be graded according to the usual severity of the attacks. Tolerance is established in the quickly recurring attacks, and changes must be made. It is not yet certain what part is played by the respective analgesic and vasodilatation actions of this group. They have robbed migraine of most of its terrors, and tended to diminish the use of morphine and its derivatives very markedly.

Caffeine is a much overrated drug. In the abortive attacks and in the morning remains of a migraine it is useful; but for a full-fledged attack it is not efficient. Similarly, bromides alone, chloral, and other widely used drugs are valuable only in mild attacks. They should be used in preference to other more potent remedies, which should be reserved for the severer attacks.

The use of aconite and cannabis indica is more restricted now that really efficient analgesics are known. Aconite is rarely called for, while cannabis indica or cannabis americana has a limited, although no less definite, place. In attacks associated with much mental depression the addition of cannabis is often useful. The often experienced inefficacy of this latter remedy is largely due to its extreme variability. Tablet preparations are usually worthless. This is equally true of the volatile nitrite preparations. Opium, or its main derivative morphine, should be used only as a last resort. It is rarely really needed.

Lying down in a quiet, darkened room—a brisk saline laxative taken as early as possible, the patient being undressed and well covered—these are essential in the severe exhausting attacks. A very hot bath often aids very materially. Cold is to be avoided.

The greatest folly of all is to treat all patients and every attack alike.

**Ophthalmoplegic Migraine.**—Attention has been called to the fact that in the ordinary attack of ophthalmic migraine there may occur various sensory or motor phenomena, among which anesthasias or paralyses are the most marked. These sensory and motor changes are extremely diverse but there is one symptom grouping which, by reason of its comparative frequency and close similarity, was set apart from others occurring in this affection and named by Charcot ophthalmoplegic migraine, in order to distinguish it from its more classical relation. It consists in a paresis or a paralysis of one or more muscles of the eye, innervated chiefly by the oculomotorius, which comes on either following or during a migraine attack. Inasmuch as oculomotor pareses or paralyses may occur from a great diversity of causes, apart from a migraine, and may appear periodically, it has been held by many that the term ophthalmoplegic migraine has no particular right to exist, but the evidence is too great to eliminate migraine as a competent producing cause for these periodic oculomotor paralyses.

**Etiology.**—Whether *heredity* plays any greater part here than in migraine in general is difficult to decide. The present attitude is to restrict the influence of heredity in migraine; certainly there is very little evidence of direct transmission of ophthalmoplegic migraine as such. Yet if there is any truth in the Spitzer hypothesis, heredity should be much in evidence in the ophthalmoplegic variety. Perhaps it is, but the cases are few and far between.

There has been much speculation concerning the central or peripheral nature of this third nerve palsy. The present view taken for migraine in general, that it is due to a disturbance in cerebral pressure, secondary to vascular modifications, is sufficient to account for the oculomotor palsies as well, in view of the location of the peripheral branches of the third nerve in regard to the cerebral vascular plexuses. In fact, the occurrence of the ophthalmoplegic type is one of the strong arguments for the general pressure hypothesis, as Spitzer has argued. If, as has been shown by several autopsies, to these considerations additional local causes be added, which increase or permanently maintain such pressure effects, the interpretation is comparatively simple. Thus, exudates, fibrous processes, swelling in the cavernous sinuses, swelling of the hypophysis, tumor formation, gummata, etc., have been found in patients suffering from periodic oculomotor paralyses associated with migraine.

It is true that some of these are to be interpreted as symptomatic migraines, in which the foreign body acts primarily as an irritant to cause the vascular disturbance, which sets free the migraine reaction, and secondarily serves as an additional cause of pressure to bring about the palsy. In a personally observed case, with basal gummata, the periodic oculomotor palsy and migraine attacks have occurred for a period extending over four or five years, usually with every menstrual period. Here were three interplaying factors, and the exact part played by each can only be inferred. The slight disturbance of menstruation, usually adjusted, in this case, by reason of the exudate, was not. A migraine was set up, the acute pressure of which, added to that of the exudate,

caused the ophthalmoplegia which had become fairly persistent in the intermigrainous interval in recent years.

**Symptoms.**—Leaving aside for the moment the atypical and symptomatic periodic oculomotor paralyses due to other causes than migraine, one finds in these patients, usually during or after a severe attack of unilateral migraine, with headache, nausea, vomiting, etc., a ptosis of the eyelid on the same side, and a loss, partial or complete, of the upward, downward, and inward movements of the eye of the same side. This eye is usually directed outward and downward, and the patient sees double. This may or may not be accompanied by sensory disturbances in the superior branch of the trigeminus.

After a variable length of time, a few days, a week or more, the paralysis disappears, usually gradually, and the patient suffers no inconvenience from the ocular palsies or the ptosis. In some individuals such palsies accompanying a migraine have come on comparatively young in life, almost with the beginning of the migraine attacks; for the majority however, they follow several years after the establishment of a migraine, in some instances as late as thirty years. In some only a very severe attack will be accompanied by the oculomotor signs, or only slight palsies; transitory ptosis may occur frequently. But in others the palsies develop with each attack of migraine and often in increasing severity. The effects may persist longer and longer between the attacks, until in a few they become permanent palsies. This type, however, often permits of other interpretations.

A double lesion can be understood, although it rarely occurs. Isolated abducens palsy has been described, also isolated trochlearis; and complete ophthalmoplegia is reported in a single case, but in view of the many contributory factors it perhaps is preferable to view such a case from another standpoint.

**Diagnosis.**—Every patient should be regarded as one suffering from something more than the migraine, until all accessory causes are excluded.

**Treatment.**—Little needs to be added to the therapy outlined under migraine. Syphilis as a cause for both a migraine and an exudate should be treated, and the Wassermann reaction utilized to clear up the diagnosis.

## NEURALGIA.

**Definition.**—A painful affection of the nerve trunk or its branches, characterized by remittent or intermittent rapid flashes of pain, with free intervals, not usually accompanied by trophic disturbances of the muscles, unless its severity limits the activities of an organ, occasionally associated with painful nerve trunks and with disturbances in the skin structures. Neuralgias are but the expression of many diverse lesions which may involve the body in general, as toxic and infectious states, a nerve trunk itself, the sensory ganglia, contiguous structures, or they may be the reflex expression of a disorder in a viscus, apparently unrelated to the nerve fibres, which are the site of the pains. Neuralgia is, therefore, to be considered solely as a symptom, a syndrome, or a purely reflex condition. Pure idiopathic neuralgia may occur solely as a psycho-

neurosis—a real hysterical conversion. They are not rare, and may involve any nerve distribution.

**Etiology.**—An extraordinary wide range of causative factors may determine mild or severe neuralgias. The most frequent are:

(a) Anemia. This is a potent factor in reducing resistance to pain and in determining the outbreak of a painful reflex disturbance.

(b) Toxins of exogenous origin, inorganic and organic, or purely endogenous toxins. Thus, poisoning by lead, mercury, arsenic, copper, especially mild cases, often causes severe neuralgias. Alcohol and tobacco are frequent organic poisons, while the toxins of many infectious disorders are especially prone to bring about neuralgias. The morphinist or cocaineist suffers severely from neuralgia as an abstinence symptom. Influenza, tonsillitis, and smallpox are familiar illustrations, while the malarial organism causes a toxemia with a pronounced aptitude to induce neuralgia. Typhoid fever, measles, gonorrhœa, possibly syphilis, and streptococcus infections are frequently accompanied by neuralgias. The toxemias of diabetes and nephritis frequently cause severe neuralgia, while those of hyper- and hypo-thyroidism are not infrequent.

(c) Inflammation of the sensory ganglia, which may be either of infectious or non-infectious nature, gives rise to some of the severest forms, as seen in herpes zoster. According to the ganglion involved, there may result an ear neuralgia—otic neuralgia, a zoster of the seventh nerve (Hunt), in one or more of its branches—zoster of intercostal ganglia, or even down to the lower extremities. Ganglion involvements of non-infectious types give rise to neuralgias such as tic douloureux, while tumors of the sensory ganglia may condition persistent and obstinate neuralgias in the distribution of the affected sensory nerves.

(d) Involvement of the nerve trunks themselves, either by mild neurotic processes, perineuritis, pressure from anatomical structures, pressure from lesions, cuts, bullet wounds, tears, tumors, aneurism, exostoses, fractures, or displacements may cause severe neuralgias, which by the advance of inflammation, the persistence of pressure or the continuance of the lesion finally show the neuritic reaction.

(e) Reflex neuralgias are numerous and puzzling. Pulmonary, cardiac, gastric, hepatic, renal, ureteric, intestinal, vesical, uterine, ovarian, prostatic, testicular, and affections of other viscera give rise to herpetic eruptions, with painful, sensitive skin areas and neuralgias; in many instances the neuralgia is not accompanied by herpes. Head's complete analysis of this class of cases is of paramount importance.<sup>1</sup> Thus, a persistent sciatica may be the reflex of a prostatic disturbance. An anemic woman may not suffer from pain, but on menstruation her referred neuralgic pains may become very severe over the tenth dorsal nerve, and pain and tenderness are frequent over the areas of the sixth dorsal (heart), seventh dorsal (stomach), and there may be occipital and mid-orbital neuralgia (Head).

(f) Psychical neuralgias are frequent. They are usually hysterical conversions, but may be found in a number of psychoneurotics and in some psychoses, mild depressed manics, and schizophrenics.

<sup>1</sup> *Brain*, xvi, 1; xvii, 339; xix, 153.



(g) Organic disease of the nervous system. General paresis, tabes dorsalis, spinal or cerebral disease, thalamus involvement, syphilitic meningomyelitis, etc., are often accompanied by neuralgic pains.

(h) Hereditary predisposition. The neuropathic constitution, hysterical personality, physical and mental overwork may be added to the conditions mentioned as responsible. Some individuals have neuralgic pains on the slightest provocation. The arthritic, gouty, rheumatic, and scrofulous may be said to be predisposed. Unknown factors thought to be related to atmospheric pressure, humidity, high electrical tension, etc., play a rôle in many of these cases.

(i) Chronic vascular disease, and especially arteriosclerosis, is a frequent cause, particularly in the aged, the senile, and the presenile. Syphilitic neuralgias are frequently conditioned by vascular disease.

(j) Last, but not least, exposure to cold is an important factor. It is not certain that all neuralgias caused by cold are not really mild types of neuritis or perineuritis, but since a neuralgia may be the expression of a mild neuritis or perineuritis, discussion of the distinction is fruitless.

**Symptoms.**—*Pain* is the main factor in neuralgia. For the most part, it is the only expression of the nerve disturbance. The character of the pain varies considerably, but in general it may be described as unilateral and paroxysmal. It is characteristic of most neuralgias that they are not primarily localized in the periphery. The pain seems to begin beneath the surface, and may then shoot out to the periphery. It may be described as biting, boring, tearing, darting, cutting, like an electric shock, like a hot iron, etc., each patient having his own pet expression. It may come and go in lightning-like flashes or throbbing pulsations, persisting for a shorter or longer time, then stopping for minutes, hours, or days, then recurring. When continuous, the pain varies considerably in its intensity. At times it is agonizing; then again it is growling and grumbling beneath the surface.

The painful area in most neuralgias conforms to the peripheral distribution of the sensory nerves. In the herpetic and referred neuralgias the root zone area is involved; this supplies an important differential in determining these types. At times strictly localized to a more or less definite spot, again the pain radiates through an entire limb, over one-half of the face, or the trunk. The radiating character of the pain in some neuralgias is very striking. It spreads out along the peripheral branches of a nerve stem. As in the trigeminal neuralgias, one may find the upper, middle, or lower branches involved; alone, in combination, or all three. A dental neuralgia may suddenly jump to other branches of the fifth nerve and the pain become general. Individual peculiarities are constantly met, in which case one theoretically assumes differences in irritability of the nerve substances, in the central gray matter.

Certain *points* seem to be foci from which the pains start. These are usually situated along the nerve trunks, and pressure upon them is often sufficient to cause an exacerbation in a mild attack, or to provoke an attack in a period of intermission. Valleix attached considerable importance to these points. They are found, according to him: (1) At the point of emergence of the nerve trunks; (2) at such situations where a

nerve trunk transverses a muscle to reach the skin; (3) at points where the nerve fibre breaks up into branches; (4) at points where the nerve becomes very superficial; and (5) at Trousseau's apophyseal points. Valleix's points play a secondary rôle in present day interpretations of neuralgias, and Romberg has given a severe critique of Valleix's claims. They are of interest particularly in the neuritic types, but are absent in other forms of neuralgic affections.

Accompanying phenomena are frequent. In some patients a sense of apprehension may precede the coming on of an attack; vague sensations of discomfort often antedate the neuralgic outbreak. Ripples of pain, like pin pricks, short twinges, etc., announce the advent of a more serious attack, or may be the sole evidence of an abortive one. Such mild phenomena are extremely frequent in certain of the so-called pre-disposed or neuralgic individuals. Some feel that they cannot live at high altitudes; others fear rain, or an east wind; a thunder storm cause others to have twinges; while, again, certain dietary indiscretions make others complain of painful twinges for days. Just what conditions are at the basis of these features is unknown, yet they are none the less real. One must always bear in mind the indubitable influence of purely mental influences; this does not make the pains any the less real but opens a view as to possible therapeutic resources.

Skin hypersensitiveness is frequent. It may precede or accompany an attack, and persist after the pain has ceased. Epicritic sensibility is mostly implicated. Light touch, a pin prick, or slight degrees of heat and cold are magnified. Deep pressure and extremes of heat and cold are usually palliative. *Anesthesia* is not infrequent following an attack of pain, and the exact topographical distribution of the sensory modifications on the skin throw considerable light on the possible causation of a neuralgia (Head). Trousseau's painful spinal point should be recalled in this connection. *Paresthesia* is very frequent, and the femoral distributions seem to show it more than others, "meralgia paresthetica."

*Motor disturbances*, either as cramp-like contractions or as paralyses, are not infrequent in accompanying conditions.

*Vasomotor* and *secretory* symptoms are frequent. The bloodvessels are frequently contracted in the early stages of a neuralgic attack, with resulting blanching and cooling of the skin. Following this a period of warmth, of redness, of free perspiration may result due to the secondary dilatation of the vessels. In many cases of trigeminal neuralgia other secretions may be modified. Crying, coryza, or salivation are not infrequent, while in general neuralgic attacks an increase in the amount of urine and of milk secreted is frequently found.

Premature graying of the hair, loss of hair, thickening of the skin, erythemata, eczema, pemphigus, herpes, thickening of the bones, and, occasionally, muscle atrophy are among the rarer trophic by-products.

During an attack irregularities of the pulse are not unusual; slowing is the rule. The pupils are frequently dilated, at times unequally.

The general physical and psychical reactions are extremely important. Loss of sleep and anorexia cause the patient to lose strength and flesh; and anxiety, irritability, and petulance are almost inevitable. Mental

depression is not infrequent, especially in severe cases of trigeminal and sciatic neuralgia. The contracting of a drug habit is not unusual.

**Course.**—This depends naturally upon the underlying condition. Many are acute and transitory, persist for three or four days, and never reappear. Such are the herpetic types. On the contrary, certain neuralgias are persistent, chronic, and show a tendency to grow worse. Tic douloureux is an example. Many reflex neuralgias run an acute recoverable course, but show a marked tendency to recurrence. The neuralgias which accompany the chronic cachexias of nephritis, carcinoma, brain or spinal cord disease usually progress in a markedly chronic manner. In those hereditarily disposed individuals the tendency to chronicity with longer and shorter periods is proverbial. Most neuralgias in which the causative factor is undiscoverable run a benign course, while the neuritic types are less amenable to treatment.

The subdivisions of neuralgic neuroses, subacute neuritic neuralgia, and chronic neuritic neuralgia offer a grouping referable to the course which has only clinical convenience to warrant it.

In the first type one finds the disorder limited more or less to the neuropath. The attacks come on without appreciable cause, or follow a nervous shock, exposure to cold, or dietary indiscretions. The pain comes on with great suddenness and usually without great violence; it comes and goes apparently without rhyme or reason, and is not accompanied by painful nerve trunks or trophic disturbances. It recovers at times, to recur at intervals of a year or years.

In the subacute neuritic neuralgic type, exposure to cold or to pressure, especially in arthritic patients, determines an attack. The attack develops gradually; the pain, at first mild and intermittent, gets worse and worse and more continuous. Finally, after a day or more the paroxysms become extreme, the intervals between marked by dull pain; Valleix's points are characteristic findings. When involving a mixed nerve, muscular atrophy or other trophic signs appear, signaling the occurrence of a neuritic process. Local cedema and herpes zoster are frequent accompaniments. This type usually commences to recover in from two to three weeks, and an ultimate recovery is to be expected. Recurrences occur, however, and leap to the third type of chronic neuritic neuralgia. This form is frequent in the aged. The history is usually that of several subacute attacks with increasing tendency to chronicity. Here the trophic disturbances in muscle and in skin are more marked. The paroxysms run a remittent course.

**Diagnosis.**—Enough has been said to emphasize the need for a searching analysis of the causative factors in every neuralgia. They are many, and presumably the most widespread diagnostic error is the overlooking of an early tabes dorsalis in young to middle aged adults. Children are not prone to neuralgias—in the narrower sense—and a neuralgic affection in childhood calls for close scrutiny.

Since unilateral pain, of special localized type, occurring in irregular attacks, is almost the sole criterion of neuralgia, it is very frequent that organic disease of a viscus will show precisely similar accompanying features. In the majority of cases the underlying organic lesion may be

detected—occasionally it remains difficult to locate. Not infrequently the diagnosis of a persistent neuralgia may be cleared up by the finding of malarial organisms in the blood, or more rarely the presence of a marked eosinophilia will call attention to trichina as the cause of an obstinate neuralgia. Syphilitic neuralgias are by no means infrequent.

The diagnosis of *myalgia* from true neuralgia is not often difficult, but occasionally, especially in the intercostal and lumbar regions, the diagnosis becomes uncertain. These neuralgic-like myalgias are usually isolated in their location, are not, as a rule, accompanied by acute exacerbations, nor are the regions usually painful on pressure. Motion, on the contrary, usually aggravates myalgia. Motion of the jaw—it may be recalled—is enough to start a severe tic douloureux.

Neuralgic pains about the jaw are at times the precursors or accompaniments of new-growths of the jaw or parotid (Gowers). Neuralgic affections of the cranial nerves are not infrequently due to intracranial growths.

*Neuritis* of a mild grade offers a specially difficult problem. As already stated, mild neuritis is a neuralgia. The question to be solved concerns the likelihood of a more severe degree of neuritis. In this case the usual signs of neuritis appear—painful swollen nerve trunks, trophic disturbances, more continuous pain, Lasègue's phenomenon, weak, flabby muscle fibres and electrical changes. New-growths pressing upon or involving the nerve trunks within or without the spinal canal, in the early stages particularly, begin their disturbance by a pure neuralgia? Minute analysis of the sensory phenomena will usually clear up the diagnosis early, although in the earliest phases at times it may be impossible.

A neuralgic affection may be one of the earliest signs of multiple sclerosis. Oppenheim has found a severe tic douloureux to have been the earliest sign of this disorder. Syringomyelia may begin as a localized neuralgia. Minute hemorrhagic lesions of the spinal cord of traumatic origin give rise to neuralgias.

In the diagnosis of *hysterical* neuralgia great caution should be exercised. Hysterical neuralgias partaking of the nature of a pseudo-neuralgia are extremely diffuse, and may react very rapidly and markedly to suggestive influences. Hysterical neuralgias are almost invariably accompanied by other hysterical conversions.

*Neurasthenic* pains need to be differentiated. The many mixed forms of neurasthenic, hypochondriacal, and hysterical origin bear their characteristic sidelights. The diagnosis of these types of neuralgia should not be lightly made, for it is not to be forgotten that these syndromes of themselves may be the reaction on the part of the nervous system to some more fundamental organic lesion. Thus, patients suffering from severe neurasthenia, with cachexia, and severe intercostal neuralgic pains may have an undiscovered carcinoma of the stomach, mediastinum, etc.

In *tabes dorsalis* the pains resemble those of neuralgia very closely, but, as a rule, have a wider range, are not localized in a peripheral nerve distribution, and are more apt to be radicular in their distribution. Pain on pressure of the nerve trunk is usually absent.

The pains, cramps, and muscular weakness of *intermittent claudication*

sometimes bear some resemblance to a severe neuralgia. Aortic aneurism gives rise to reflex neuralgic pains, usually very severe. Aneurisms in other regions are to be carefully excluded.

In reflex neuralgias the use of cocaine or other local anesthetic may determine by exclusion, the site of the original lesion. An orthoform suppository pressed well against the prostrate has been known to relieve a severe sciatic neuralgia. Tumors of the pelvis frequently give rise to sciatic and crural neuralgias, and persistent neuralgic pains of the knee are often a reflex from hip-joint disorder.

**Prognosis.**—This is conditioned by the pathological process that is responsible. The more chronic of the neuralgias, which in years gone by tended to bring about chronic invalidism or inveterate drug habits, have ceased to have such a sinister import by means of a better understanding of the underlying conditions, and by a much more resourceful therapy. The younger and stronger the individual, and the less the tendency to hereditary disposition, the better the prognosis in those neuralgias which are idiopathic, as well as those due to alcohol, lead, etc. In the more chronic forms, not due to removable conditions, the prognosis is bad.

**Treatment.**<sup>1</sup>—The chief indications are to quiet the pain and root out the cause. A painstaking study of the history and an exhaustive physical examination are necessary in all cases. The therapy will vary, therefore, widely if the cause be ascertained; a course of quinine will cure one patient, a surgical operation may be called for in another.

Taking up the general therapeutic indications, the *analgesics* which have proved serviceable may be discussed first. From the true therapeutic standpoint they are solely palliative. Many cases of severe neuralgic pain may be temporarily subdued by the synthetic analgesics and the dangers of a morphine habit averted. Phenacetin, acetanilide, antipyrine, aspirin, pyramidon, lactophenin, and phenocoll are among those that have proved valuable. New ones are being added, and among them some are certain to be of value. The salicylic acid group combinations are at times useful, especially in the milder cases and in patients with arthritic tendencies. In influenza and tonsillitis neuralgias the salicylates are useful. Combinations of these with hypnotics, such as chloral, paraldehyde, sulphonal, trional, or veronal, are useful in procuring sleep, and thus prevent the reduction of the patient's resistance.

If any of the opium group be necessary, it is better to give such in sufficient doses. Usually smaller doses may be given when combined with the analgesics mentioned. Aspirin, gr. vii (0.5 gram), codeine, gr.  $\frac{1}{3}$  (0.02 gram), and trional, gr. vii (0.5 gram), for instance, is a useful combination in mild cases. In chronic neuralgic pains morphine is to be avoided as long as possible. This does not apply to a very old patient, or one in whom the neuralgia is simply the expression of some chronic incurable disorder—carcinoma, for example. The gradual immunity acquired, with the need for larger doses, and the pernicious effects of a habit apply to all the members of the opium group.

<sup>1</sup> Schmidt, in *Modern Treatment of Nervous and Mental Disease*, edited by White and Jelliffe.

It is doubtful if chloroform or ether is to be recommended by inhalation in severe neuralgias. They are useful temporarily in great crises.

Other drugs are quinine, which in combination with the salicylates is specially valuable; arsenic, which is serviceable in the neuralgias due to anemia, especially in combination with iron. Atropine and aconitine were used widely before the days of the antipyretic analgesics. Their definitely poisonous qualities have driven them into the background. The unreliability of *Cannabis indica* has done the same for this otherwise useful analgesic. The iodides are called for in the syphilitic neuralgias, and are useful in many neuritic neuralgias.

*Counterirritation* is of great service in most cases of severe neuralgia, especially after the acute onset is over. The Paquelin cautery is the best means; mustard paste, cantharides, turpentine, chloroform, ether, and acupuncture all have their place. Local freezing may be carried out by ethyl chloride, methyl chloride, ether, or other volatile substances. Menthol, or other similar derivatives may be used for mild neuralgic pains to advantage. Local applications of heat are grateful and valuable. Hot water bags, hot sand, electrical pads, etc., may be utilized. General or local hot water baths, or hot air baths (baking), are at times desirable.

Direct applications of local analgesics, either to the nerve trunk or within the spinal canal, are valuable in many obstinate deep-seated neuralgias, especially of medullary origin. Cocaine, tropacocaine, eucaine are all useful given by the Corning or Quincke method.

General hygienic treatment is imperative. A generous diet, full sleep, healthful occupation, and freedom from mental worry are essential. Cod-liver oil, nitrogenous diet, with iron, arsenic, strychnine, calcium salts, are indicated. Faddy dietaries should be avoided. Even in arthritic neuralgias it is doubtful if meat does any particular harm when not taken to excess. Alcoholic beverages are to be denied.

Hydrotherapy may be added to other hygienic means, if it proves invigorating and causes an increase in appetite and sleep.

Climatic changes are often advisable. Low-lying, damp, and humid atmospheric conditions seem least desirable. The general stimulus that comes from a drier, higher atmosphere, even if colder, works to the general advantage, even if not directly valuable for the relief of pain.

Electrotherapy, when well managed and properly selected, is of great value in some neuralgias. In general, Leduc's modifications of the d'Arsonval's rapidly interrupted current offer the readiest and most widely applicable form of electrical current for the relief of neuralgic pain. Such treatment is purely palliative and is pernicious if it leads one to overlook causal factors. Most of the cures due to galvanism are psychical. Some intractable neuralgias need a psychoanalytic cure.

*Surgical* intervention is called for in all cases in which pressure is demonstrable and the cause removable. Tumors and new-growths, involving or pressing upon nerve structures, if removable, should be taken away. Surgical interference may be of radical service in many of the reflex neuralgias of obscure origin probably related to visceral ptoses. Nerve stretching needs mention mostly to be condemned.

## SPECIAL LOCALIZED FORMS OF NEURALGIA.

While any nerve fibre in the body may become painful, there are certain regions which show a greater tendency to involvement than others. Bernhardt has collected the statistics of localized distribution in some 685 cases, with the following results: Trigeminal, 124; occipital, 42; brachial, 108; intercostal, 45; lumbo-abdominal, 12; crural, 25; obturator, 2; sciatic, 303; anterior femoral, 11; Achilles, 3; tarsalgia, 4; metatarsalgia, 4; and coccygeal, 2. In 613 cases collected in the clinic of Dr. M. Allen Starr during four years (1902 to 1906) the distribution was as follows: Trigeminal, 315; occipital, 28; brachial, 31; ulnar, 1; intercostal, 19; lumbo-abdominal, 19; crural, 2; sciatic, 194; coccygeal, 1; peroneal, 2; and plantar, 1.

Thus, with the New York population the affections of the three branches of the fifth stand first, the sciatic next, while the brachial, occipital, intercostal, and lumbo-abdominal follow. Bernhardt, Lachnit, and Conrad have found sciatica to be the commonest type, with trigeminal neuralgias second. Eulenberg has found, with the author, that the trigeminal affections have been more frequent. In the Vanderbilt Clinic (New York) the trigeminal neuralgias have been more common in women, while the sciaticas have predominated very markedly in men.

**Trigeminal Neuralgia.**—Neuralgias of the branches of the fifth nerve are among the commonest of all the neuralgias. The inferior and superior branches preponderate in frequency of involvement. Most frequently these neuralgic pains are due to some affection of one of the branches. Inflamed teeth play a minor rôle. Affections of the ears, the eyes, iritis, cyclitis, iridocyclitis, the skin of the face or head, inflammation within the accessory sinuses of the nose, forehead, antrum, mastoid, all of these may produce diffuse neuralgic pains, at times clearly separable from a simple neuralgia of the fifth, at other times not. The characteristic type of trigeminal neuralgia is *tic douloureux*; trigeminal herpetic neuralgia, involving one or more branches, is not uncommon.

Cold and wet are very important agents in facial neuralgia. In certain countries, notably England and the north of Germany, facial neuralgias from this cause are extremely common; they seem to be much less frequent in the United States, and notably so in southern countries.

*Tabes dorsalis*, or multiple sclerosis, may make its first appearance as a trigeminal neuralgia.

Neuralgia of the superior branch is seen more commonly by physicians, although the dental branches are involved much more frequently. These patients go to dentists, and hence do not enter into medical statistics. This is a reason why it is incorrectly stated, by most writers, that the superior branches of the fifth are most often involved. For the most part the milder types of facial neuralgia are induced by irritation of some of the terminal filaments, while in the severe form, *tic douloureux*, which is the more classic, a lesion of the Gasserian ganglion is uniformly present. Mild cases of *tic douloureux* may be indistinguishable clinically from other types of facial neuralgic pain.

It would be desirable to restrict this term to a definite and, if possible,

limited type of neuralgia of the fifth nerve, particularly to the form due to changes of a chronic degenerative nature occurring in the Gasserian ganglion. This is not yet possible, and clinically the neurotic and peripheral cases are either not at all separable from the ganglion cases, or with considerable difficulty.

It affects one side of the face only, as a general rule. In certain generalized neuralgias, such as those due to diabetes or nephritis, both fifth nerves may become involved. In the majority of cases selection occurs among the branches, one or two being involved, rarely all three; the ophthalmic branch the oftenest, the inferior maxillary the least.

The more classical tic douloureux neuralgias are characterized by the extreme severity of the pain, usually preceded by paresthetic prodromata, and widely accompanied by sympathetic or irradiating pains in other branches than the one chiefly involved, or in other nerves. The pain may be paroxysmal or continuous, with marked exacerbations. They have been compared to the piercing pains of a sharp knife or the burning of a red-hot wire. The patient remains for a shorter or longer period, a few minutes to several hours, under the grip of the pain, unable to move a muscle of the face or fearful of stirring, lest a spasm more fearful than the others should occur. The slightest touch is avoided, even the air pressure of a suddenly closed door may bring about an exacerbation. The longer attacks are rarely as vicious as the shorter ones.

Valleix's points are relatively constant. In the ophthalmic involvement the sore parts are found above the supra-orbital notch, at the external angle of the upper lid, the upper outer aspect of the nose, and the globe of the eye; in the superior maxillary branch the inferior orbital notch is the chief point of pain; the malar bone, and opposite the last upper molar are other less frequently found points, while the outer angle of the mouth and the roof of the mouth are rarely their site. In the inferior maxillary distribution the points are chiefly just in front of the auditory canal, the site of the tongue, the border of the chin, and Trousseau's points over the first and second cervical vertebræ.

Vasomotor and secretory disturbances are usual. The skin is, as a rule, hot and swollen, occasionally pale and frigid; tears, nasal secretions, and saliva flow in abundance. The eyelids may be swollen, the conjunctiva reddened, to the point of ulceration at times; within the nose and mouth extravasations occur, and ulcers are not uncommon. Herpetic attacks are also not infrequent, and in some of these attacks grave injury to the eye structures may take place. Glaucoma is one of the severe complications. Other trophic disturbances have been noted, such as skin eruptions, acne, erysipelatous reddening, graying of hair, and blackening of the tongue. In continued cases hemiatrophy is known to occur. Changes in the sense of taste, of touch, of hearing, are at times present. Photophobia is frequent, while diminution in the visual fields and accommodation cramps are noted. Gellé has described a neuralgic deafness, due possibly to labyrinthine effusion in protracted cases.

Severe mental disturbances, amounting at times to hallucinatory confusion, may be present, and profound states of depression, which have resulted in suicide, in isolated cases, are to be guarded against.



The motor disturbances consist in convulsive movements of the facial muscles (convulsive tics, spasmodic epileptiform neuralgia, Trousseau), sudden forced closing of the eyelids, drawing of the mouth to one side, or sudden turning of the head. At times the convulsive movements extend to the arms. Paralytic phenomena in the third nerve are noted. The general physical disturbances noted are prone to occur in this type.

**Course.**—In the majority of cases the attacks appear in series and attain a periodicity which comes to be dreaded by the sufferer. The free intervals usually become shorter and shorter; but many patients may have only one attack a year, especially in cold weather, or even at longer intervals. A single attack may last a few days, or in the severe forms several weeks, the patient not being free from pain day or night, save under the influence of morphine. Some patients have a few attacks in a lifetime, others are not free from the disease for years. The severer convulsive forms are prone to occur late in life, when resistance is low.

**Diagnosis.**—Ordinarily the classical form of tic douloureux is recognized without difficulty. Patients have had all their teeth extracted, however, under the mistaken diagnosis of dental disease, while some intractable facial neuralgias have been cured by proper attention to diseased teeth. Aneurism of the carotid, tumors pressing upon the nerve or upon the Gasserian ganglion, may be difficult to determine as the exciting cause. These, however, are usually accompanied by accessory symptoms, palsies, eye-ground changes, aneurismal murmurs, pain within the head, cerebellar syndromes, ear pains, etc.

**Treatment.**—It is as essential to endeavor to find and treat the cause of a facial neuralgia as for neuralgia in general. The various remedies given under the heading of neuralgia may be tried, and as malarial neuralgias are very frequently facial, energetic quinine therapy should be given; the absence of blood findings is not contra-indicative, especially in malarious regions. Arsenic is a useful adjuvant in these patients, and also valuable in non-malarial neuralgias. Gelsemium, the tincture in 10 minim doses, gradually ascending, aconitine in doses of  $\frac{1}{500}$  grain, Cannabis indica (fresh) in doses of  $\frac{1}{4}$  to  $\frac{1}{2}$  grain, are reputed as specially valuable in the facial cases. Any of the analgesic antipyretics may suit individual cases, and avoid the use of morphine, which alone is reliable in many severe cases.

Injection methods have been tried for years. Osmic acid was used which set up a degenerative process, and in many instances relief was obtained, but regeneration took place even when the injections were practised within the main trunks. Schlosser has perfected a method of injecting alcohol within the substance of the Gasserian ganglion, which has given excellent results. The chief feature in the deep alcohol injection method is the introduction of a long, dull, cutting needle into the foramen ovale and there injecting *in situ* the branches of the trigeminus. Narcosis is not necessary; 80 per cent. alcohol is used. In three to four hours following the injection pain is relieved, and two or three more injections are given within a week to complete the treatment. Immediately following the injections, there is a marked anesthesia on one side of the anterior part of the head, including the nostril, palate, and one-

half the tongue; a slight paralysis of the muscles of mastication, which usually disappears in a few hours; a degenerative process is set up in the nerve trunk, which is recoverable, and general sensibility usually returns, but the pain is absent. Relief extending over a year in a number of cases is reported by numerous observers. Some patients have been relieved for four or five years. Edema of the posterior eye structures and hemorrhage are among the discomforts and even dangers of the operation, especially in the use of the intra-orbital methods devised by French operators. Repeated injections delay the recurrences more and more, and thus the patients may be held immune to the pain. Numerous modifications and later methods have been devised.

As regards surgical means, three procedures have been seriously advocated. The first and earliest consisted of peripheral section. Section of the fifth may be employed to advantage in those cases in which the disease is undoubtedly peripheral. As modified by more recent procedures, the older objection that regeneration takes place is partly done away with, and in selected cases peripheral section may be preferred to the more severe and serious operations.

Rose,<sup>1</sup> in 1890, first successfully extirpated the ganglion for tic douloureux. Horsley, Hartley, and Krause further perfected the operation, and the modified Hartley-Krause operation by the temporal route has been largely the method of choice. Cushing's modifications are of lasting value. The operation still remains one of much difficulty and seriousness. Recurrences are known even with this method.

Van Gehuchten,<sup>2</sup> in 1903, suggested what he termed the physiological section of the roots of the Gasserian ganglion by tearing. His extensive studies on regeneration in nerve fibres showed him that the central side of the ganglion must be attacked if the problem of recurrence was to be solved, and, furthermore, his method took into account the small number of intracranial cases not reached by the more peripheral methods. Spiller, as early as 1898, had suggested the surgical expedient of cutting the sensory root, which he claims is safer than the operation of tearing. The method of division of the sensory root promises to be one of the most valuable surgical procedures thus far devised.

**Cervico-occipital Neuralgia.**—This occurs in the distribution of the sensory nerves of the cervical plexus, consisting chiefly of the occipitalis major, the occipitalis minor, auricularis magnus, cervicalis superior, supra-clavicularis and phrenic. Neuralgia in this region seems rare.

**Etiology.**—The several causes of neuralgia are operative here, and need not be repeated. Special determining features seem to be the carrying of heavy weights on the shoulders (a more frequent cause seen in brachial neuralgias), arthritis deformans of the upper cervical vertebræ, caries, syphilis, tuberculosis, tumors, pachymeningitis, falls and blows wrenching the cervical vertebræ, enlargement of the cervical lymphatics, and aneurisms of the vertebral artery. Psychological neuralgias in this general region are infrequent.

The pains occupy the regions mentioned, being particularly localized

<sup>1</sup> *Lancet*, 1892, i, 295.

<sup>2</sup> *La Neuraxe*, 1903, v, 201.

in the neck, below the occiput, and running up to the vertex, occasionally behind the ears. The Valleix point found most frequently is the occipital point between the mastoid apophysis and the first cervical vertebra; points between the sternomastoid and trapezius (cervical), the anterior border of the mastoid, and the middle of the ear are of less frequent occurrence. The pain is very frequently bilateral. Dull pain on pressure, with tender skin, is usual as an interparoxysmal occurrence. This tends to make the sufferer hold his head in a stiff position, which in time may cause a characteristic attitude. The tenderness may be so acute that ruffling of the hair will start a paroxysm. Graying of the hair, loss of hair, with other trophic signs, may be present. Sudden pulling back of the head is an occasional symptom.

**Diaphragmatic Neuralgia.**—This form of neuralgia, also known as phrenic neuralgia is of rare occurrence. The pain is usually present near the free border of the ribs, or within the chest, between the ribs, occasionally as high as the chin and in the neck, beneath the clavicle, and in the scalenus anticus muscle. Trousseau's points are located over the second to the fifth cervical vertebra. The pain frequently runs down the arm, especially in certain complex cases of mixed brachial neuralgia. Breathing may be seriously interfered with, the breath coming fast and short; longer excursions of the diaphragm are impossible. It is a common experience to have a sharp, short stitch in the side, with inability to breathe for fear of pain. In the majority the pain is in the left side. Anemia, affections of mediastinum, heart, and pericardium, and aneurism of the aorta are the most frequent attending features. An intractable phrenic neuralgia may complicate an exophthalmic goitre, or be present in carcinoma of the neck region.

**Brachial Neuralgia.**—In this general form the components of the brachial plexus, from the four lower cervical, or some of its filaments, and first dorsal roots, are those involved. The chief nerves carrying sensations from the skin area of the arms and shoulders are the circumflex, radial, internal cutaneous, and musculocutaneous. These enter, for the most part, the upper and middle cords of the plexus. In the majority of cases the pains of brachial neuralgia are located in the upper arm and about the shoulder, *i. e.*, in the area of the circumflex, radial, musculocutaneous, and internal cutaneous nerves.

Bernhardt's statistics show that men are more frequently affected than women, but the reverse shows true in the figures of other observers (Romberg, Erb). More women come to the Vanderbilt Clinic for brachial neuralgia than men, and in most instances it seems that excessive sweeping is the attributed cause. In piano players neuralgias in this area are frequent. Perhaps these should be relegated to the occupation neuroses with the pains of hair dressing, skirt carrying, telegraphy, writing, etc. At any rate, arm and shoulder pains are frequent in their mild grades at least, and very variable. The usual causative factors all come into play here. The neuropathic constitution is put in the foreground by Oppenheim; Bernhardt lays considerable stress upon the importance of bone injuries with callus formation in the formation of many arm neuralgias. Small punctured wounds about the wrist, forearm,

and arm are responsible for many symptomatic neuralgias. Vertebral disease, tumors, aneurisms, syringomyelia, multiple sclerosis, tabes, and syphilis are causes. Cervical rib should not be overlooked.

**Symptoms.**—Cervicobrachial neuralgias are extremely variable in distribution, extent, and severity. The onset is usually sudden, especially in those patients in whom an antecedent history of exposure to cold and wet is obtainable (motormen, policemen, etc.); at times the beginning is preceded by twinges and slight distress. The pains in brachial neuralgia are less apt to be the sharp shooting variety so dreaded in *tic douloureux*, but sudden accession of sharp pains, varying in their intensity, are frequent. As with most neuralgic pains, movement increases them. Toward evening the pains are apt to increase and the patient, although obtaining relief by lying down, rarely sleeps well. Soreness of the skin, slight swelling, and general reduction in tone are the usual accompaniments. With increasing disuse slight atrophy is common, and swelling is usual. The tendon reflexes are usually more irritable and active. More marked atrophy paresis with vasomotor trophic symptoms and altered tendon reflexes indicate a definite neuritic process. Herpetic eruptions occur with non-infectious, as well as infectious involvement of the sensory ganglia.

Tender points are very variable. They are most frequently in the middle of the back; about the level of the second or third dorsal there is usually a sore Trousseau point. Gowers noted that the inferior ulnar point in front of the wrist is the commonest sore point. Babinski has called particular attention to a radial neuralgia due to a mild or severe neuritis of the radial. The pains occupy the posterior portion of the arm, and are unusually severe. Neuritic changes are not infrequent.

**Diagnosis.**—Particular care is needed in excluding affections of the spinal cord, meninges, and vertebræ, as well as angina pectoris. Disease of the joints and bones should be excluded at the outset, although it may be impossible in some cases of periostitis. In tumors and other organic affections of the cord the painful points are usually absent, but the earliest and only symptom of spinal cord tumor, intramedullary or extramedullary, may be a brachial neuralgia. In tabes the pains are apt to be bilateral.

The occupation neuroses involving the arm and shoulder are many. The history of the protracted exercise of certain groups of muscles is usually sufficient to identify the proper cause for the neuralgic pains. Occupation neuralgias, like neuritic neuralgias, are neuralgias none the less, the sole diagnostic question arising as to the cause, and through this the proper mode of therapeutic attack and the probable outcome. Alcoholic neuritis in its mild grades offers particular embarrassments. Lead poisoning is to be borne especially in mind, while diabetes is of importance. Brachial psychalgia is a possibility, but the diagnosis must be made with extreme caution after exclusion of organic factors.

**Treatment.**—Rest is a necessity, and is primarily insured by the use of a sling. The diagnosis of the cause being assured, treatment should be begun to remove it, either by medical or surgical means. Nerve suturing for injury has come to occupy an important part, and is usually

attended with good results, even after long periods of loss of function. In the early stages active mechanical treatment is to be avoided. Hot applications are useful in most acute neuralgias. In later phases massage, particularly the Nügeli movements, are valuable. The Leduc rapidly alternating currents are frequently palliative. Salicylates (especially in analgesic combinations), iodides, quinine, arsenic, and large doses of strychnine are of value.

**Intercostal Neuralgia.**—The twelve dorsal nerves constitute the plexus involved, although the upper series, especially of the left side, are most frequently concerned. Bernhardt says that the site of election is mostly from the fifth to the ninth. Since the dorsal nerves divide into internal and external branches the site of the neuralgia may be on the surface or within (pleurodynia, etc.). The two upper nerves send branches to the internal surface of the arm, and pain is occasionally felt there. The abdominal involvements are rarer, and may extend down to the genitals.

The pains are usually less severe than in other regions, although their sharp, sticking character distresses the chest movements, especially since all motion tends to aggravate them. Tender points are found at the site of perforations near the spine. Skin hyperesthesia is extreme at times. Herpetic neuritic neuralgias are relatively common in this distribution. *Mammary neuralgia* or *mastodynia*, which is frequent in the later phases of nursing, and in some women at the menstrual epoch, is a special form. The pain is usually deep within the gland, and may be accompanied by a slightly increased secretion. The whole skin may be sensitive, especially the nipple, when the superficial nerves are mostly involved.

Among the causes to be diagnosed may be costal caries, affections of the spinal cord and meninges, disorders of the pleura, particularly carcinoma and tuberculosis, tabes, aortic aneurism, dilatation of the stomach, carcinoma of the liver, angina pectoris, pericarditis, trauma, etc.

**Treatment.**—Local applications are useful, especially the ethyl chloride spray. Blisters are efficacious. Bandaging affords marked relief.

**Lumbar Plexus Neuralgias.**—These are most conveniently arranged as (1) lumbo-abdominal, (2) ilio-scrotal or testicular, (3) crural, (4) femoral, and (5) obturator, involving in each case certain of the branches of this plexus. Mixed and indeterminate forms are not infrequent.

1. **Lumbo-Abdominal.**—These occupy the lower half of the trunk, and are extremely variable. The chief nerves involved are the iliohypogastric and its branches, the inguinal, and genitocrural. Strict localization to one trunk is rare, and men are more frequently affected than women. The chief causes in addition to those of general moment are local inflammatory conditions or new-growths involving the plexus or some of its branches. The pains are usually unilateral, occasionally bilateral, involve the region of the back below the ribs, the gluteal region, the abdominal and inguinal areas, the scrotum, or the labia. The chief Valleix's points are over the lumbar vertebrae, the hip or iliac point, hypogastric point, and the scrotal point. Lumbo-abdominal pains are usually accompanied by intercostal pains above or thigh pains below.

2. **Testicular Neuralgia.**—Astley Cooper termed this neuralgia the “irritable testicle.” The pains are usually unilateral, pass into the testicle, which may be swollen and tender to the touch. The pain not infrequently passes into the leg and back, and the patient may have an attack of vomiting. Bernhardt notes that the pain may be so intense as to cause the patient to seek castration. The affection is an obstinate one, and is not helped, as a rule, by removal of the testicle. Diagnosis involves a rigid exclusion of organic disorder of the testicle, although many affections (gonorrhœa, tuberculosis, chronic prostatitis, etc.) are not infrequently accompanied by persistent neuralgic pains.

3. **Crural Neuralgia.**—The crural or femoral nerve is here implicated. The pain extends in the upper front and inner side of the thigh, to the knee, and further through the saphenous distribution to the ankle and inner aspect of foot, extending as far as the big toe. It is almost entirely confined to men, and shows considerable variability as to the branch involved. It not infrequently accompanies a sciatica. Special etiological features are found in fecal impaction, or even chronic constipation, disease of the hip or knee bones, enlargement of the inguinal glands, aneurism of the iliac artery. Charcot called attention to the frequent association of crural neuralgia and diabetes. Spinal arthritis and tabes are obscure causes.

Movements of the thigh usually are painful, and the patient comes to bend his body forward in a strained position. The painful points of greatest frequency are just below Poupart’s ligament, just within the inner condyle, over the malleolus, inner side of the instep, and one over the great toe. Neuro-atrophic changes usually occur in the quadriceps, but the patellar reflex is rarely affected, save when a definite neuritis is present. Herpes, reddening, and hyperesthesia are not infrequent. In the diagnosis, disease of the inguinal vessels is to be looked for, as well as intrapelvic disorders, new-growths, etc. Crural neuralgias have a fairly good prognosis.

4. **Femoral Neuralgia.**—Here the cutaneous femoris lateralis, arising higher up in the pelvis, is involved. The pain is felt in the upper and outer aspects of the thigh, extending to the knee. A painful point over the anterior spinous process of the ilium is usual. Paresthesia in the distribution of this nerve has been extensively studied (*Meralgia parasthetica*.) The relation of the pressure of corsets has been pointed out, and much sitting is frequently associated with this neuralgia. The prognosis is favorable.

5. **Obturator Neuralgia.**—Lesions of this nerve are fairly constant as a result of the pressure of the intestinal loops of a hernia. The pain is located in the inner side of the thigh, and is accompanied by a feeling of stiffness, creepy, crawly feelings of the skin, and inability to bring the thigh toward the middle line of the body.

**Neuralgias of the Pudendal Plexus.**—A large number of neuralgias of the general plexus are recorded. The median hemorrhoidal branches, distributed to the rectum, bladder, and vagina, the inferior branches to the anus, and the pudendal nerve supplying the testicular sac, the labia, penis, urethra, and clitoris, are the chief nerves involved. The general

terms, spermatic neuralgia, anal neuralgia, perineal neuralgia, rectal neuralgia, vesical neuralgia or cystalgia, urethralgia, prostatic neuralgia, penis neuralgia, irritable uterus, ovarian neuralgia, are utilized to describe these different affections. These neuralgias are very rare, but often very obstinate. Spermatic neuralgias are among the most frequent, and not infrequently accompanied by priapism, perhaps ejaculation.

Since the advent of bicycle riding neuralgias of this general region have been on the increase. The ovarian neuralgias are complex, and more often come within the domain of the gynecologist, as structural defects are usually the underlying causes. Localized herpetic eruptions accompany neuralgias of this plexus. Lesions of the cauda equina are to be carefully excluded in neuralgias of this region.

**Neuralgias of Coccygeal Plexus.**—Coccygodynia, painful coccyx, is a not infrequent disorder in women, especially in multiparæ and in the badly constipated. Trauma and caries are frequent causes. The hysterical coccyx is not infrequent, and referred coccygeal pains are common. The pain is so intense at times that defecation is rendered impossible; the patient cannot sit, and a grave neurasthenic condition supervenes. The medicolegal significance of coccygodynia is real, appearing frequently as a local symptom of a general traumatic neurosis. Surgeons frequently lay considerable stress on a freely movable coccyx in accident litigation. A just estimate of the true bearing of an injury to the coccyx can only be arrived at by a careful survey of all of the factors of the particular case.

Local treatment is seldom efficacious save in the truly neuralgic types. Resection is rarely a justifiable procedure. Nägeli's massage movements are particularly efficacious in remedying the mechanical obstructions often conducive to persistent constipation.

## OCCUPATION NEUROSES.

**Synonyms.**—Professional neuroses; professional cramps; *crampes fonctionnelles et professionnelles*; *spasmes professionnelles*; *impotence professionnelles*; *Beschäftigungsneurosen*; *Beschäftigungskrämpfe*.

**Definition.**—These are abnormal nervous modifications of voluntary muscular coördinations involving certain muscle groups, which are subjected at frequent intervals, by reason of a definite occupation, to fine, complicated, and coördinated movements. They are known under a great variety of names, such as writer's cramp, pianist's cramp, violinist's wrist, telegrapher's cramp, milker's cramp, tennis elbow, baseball pitcher's glass arm, dancer's cramp, etc. For the most part, they consist of tonic contractions, with or without pain, intermingled at times with clonic shocks or tremors. The associated movements become more and more difficult to carry out, until a motor paresis terminates the attempt, the muscles all being tied up, as it were, into a painful tonic contraction. Muscular atrophy may appear later. A great diversity of objective manifestations is to be observed. In general, two features are characteristic. The spasms occur in muscle groups which carry out some habitual or professional synergistic act, and the

cramp occurs only when the act itself, *i. e.*, the bringing together of the entire willed mechanism, is performed. When the individual muscles are acting independently in some other less frequently performed movement no such cramp occurs. The implication of mental impulses is highly probable.

The recent monographs of most value are those by Remak,<sup>1</sup> 1894, Bernhardt,<sup>2</sup> 1898, and the article on Crampes Professionnelles by Meige.<sup>3</sup> Chapters by Gowers, Oppenheim, and Gasset may also be cited. Thomas has written an important monograph.<sup>4</sup>

**Etiology.**—The chief factor is the frequent and continuous use of certain muscular movements in an occupation beyond the natural capacity of the individual to stand the strain. Added to this are usually certain minor or major features which determine why that particular individual should develop the neurosis. In many instances an antecedent pressure neuritis reduces the capacity of certain groups of muscles involved in a coördination, and this throws increased stress on others. In others, faulty methods of adjustment, as in writing in a cramped position, or with improper light, may prove a determining factor for one individual. Mention may also be made of definite physical ills which contribute to reduce nervous tone. These are not unimportant; anemia, lead poisoning, alcoholism, excessive use of tobacco, venery, diabetes, tuberculosis, the beginnings of serious nervous troubles, paresis, tabes, etc., may be distal causes. Accident, with strain to muscle, tendon, or joint, may be the straw which breaks down the adjustment. Mental causes may show in others; anxiety, worry, financial stress may cause persistent insomnia, which may contribute to an occupation neurosis. The influence of *heredity* is difficult to trace, for no family is free from some disorder or disease, and the evidence is too flimsy to build upon.

In well-developed cases, more particularly since there is such a mental element, it is not unnatural that *psychical* causation factors should be postulated. Duchenne advocated the view that mental factors were important. Meige follows more closely the original psychic germ sown by Duchenne, and places the mental factors in the foreground. He believes that the occupation neuroses are essentially allied to the tics, resembling them not only in their objective features, but also in their mode of appearance and evolution in the disposed. In the vast majority of these cases, he holds that the motor trouble appears very distinctly as a consequence of the mental trouble.

The multiplicity of forms described—contracted, spasmodic, ataxic, tremulous, paralytic, etc.—all arise on this common mental basis; they may be found isolated in one patient, or may exist in various combinations. All distinctions between professional and functional spasms, cramps, etc., Meige holds, are arbitrary. Psycho-analysis has shown the mental factors even more clearly.

<sup>1</sup> Eulenberg's *Real-Encyclopädie der gesamten Heilkunde*, new edition, 1909.

<sup>2</sup> *Die Erkrankungen der peripherischen Nerven*, Tiel, ii.

<sup>3</sup> Bouchard's and Brissaud's *Traité de Médecine*, second edition, 1905.

<sup>4</sup> *Modern Treatment of Nervous and Mental Diseases*, White and Jelliffe, vol. i.



**Occurrence.**—Those coördinated actions of most frequent performance will naturally present themselves the more often as disordered. The practitioner in the large city will practically see more writer's cramp than other types. The practitioner of rural districts will see milker's palsies. The medical man in miners' camps will be called more often to treat the pick cramp and miner's nystagmus. In every large city there are workers in certain occupations in which fine manual work is carried on for long hours, under adverse circumstances. Many of these workers develop very severe grades of occupation neuroses, but their study has not been at all systematic.

Many writers say that the disorder is more common in men. This statement is made largely on the evidence afforded by the sufferers from writer's cramp, but since women have taken up telegraphy, stenography, and typewriting, and are engaged so widely in the manufacture of small articles for the trimming of clothing, hats, etc., it would seem that they now show more forms of occupation cramps than men do. Gowers' statement regarding stenographers not developing writer's cramp is erroneous. A limited experience has revealed at least a dozen stenographers, male and female, who have had to give up their work because of occupation cramps.

The *age* of onset is usually after twenty years. Making a combination of the reported cases (194) of Berger, Poore, Gowers, Remak, and Bernhardt, the distribution is as follows: Ten to twenty years, 4; twenty to thirty years, 67; thirty to forty years, 63; forty to fifty years, 45; fifty to sixty years, 10; and sixty to seventy years, 5. To which may be added 46 personal cases: Ten to twenty years, 4; twenty to thirty years, 12; thirty to forty years, 26; and forty to fifty years, 4.

**Occupation.**—Practically every known occupation has its neurosis. To describe them in detail is impossible. Even new sports introduce new occupation neuroses, such as the golfer's "wrist," the tennis "elbow," baseball "glass arm," etc. Blacksmiths, coopers, carpenters, masons, ironers, tailors, and washwomen show the peripheral neuralgic types, usually implying pressure. Among cigar rollers, those who crochet, knit, sew, or cut, rinsers, cabinetmakers, planers, machinists, locksmiths, weavers, chemists (holding beakers and test-tubes), players of musical instruments, occupation neuralgias, which may never advance to occupation palsies, are known.

**Symptoms.**—Irrespective of the occupation, the development of a professional neurosis seems to proceed along fairly definite lines. Individual variations are found, however, and certain persons, by reason of unknown factors, do not travel as far as others in their journey to the completed stages. It is for this reason that the etiological features upon which Meige lays so much stress may enter into the mastery of the situation. In the majority mild or severe occupation neuralgias are first observed; many patients never have anything else, and a few never have these. Occupation palsies, or impotence, as the French call it, present another phase; these may represent the results of a general or local asthenia and bear no relation to the fully developed neurosis such as is seen in a typical writer's cramp.

In those forms in which a general asthenia is at the basis of the motor loss, anemia, cachexia, alcoholism, diabetes, tuberculosis, etc., are often present. Thus, in many occupations, which call for localized, restricted, muscular adaptations involving considerable muscular power, as in cigar rollers, engravers, and glassblowers, localized atrophies develop, sometimes as a result of pressure on an exposed nerve, sometimes from other causes. These pressure palsies and pressure atrophies should receive very careful scrutiny. Some of the more characteristic are seen in waiters, in butchers who carry heavy pieces of meat over the shoulder, in others who carry packs, in steel workers, in steersmen who use the arm to push the rudder, in polemen (as seen in the European canals) who push the boat by their long poles against an elbow. Potato peelers, vegetable preparers, drummers, polishers, planers, etc., all show such muscle pareses, accompanied usually by neuralgic manifestations.

For many of those patients in whom *atrophies* develop, a local cause may be found. Some nerve stem is caught between the structures of the hand and a surface pressed upon; in rare cases perhaps a nerve may suffer from pressure of tendons when such tendons are constantly on the stretch. These local atrophies, when seen apart from the more definite development of muscular incoördinations in a complicated motor act, play a minor rôle in the true occupation neuroses, and are mentioned solely because such atrophies are sometimes contributory factors in the production of the sense of fatigue, and are the cause of paretic phenomena in others. In the sense that these show pure peripheral origins they differ from the typical cases of occupation neuroses (writer's cramp), although it seems that transitional stages are observed between the peripheral cramp-like states and the centrally induced phenomena.

Remak has called attention to the cramps of sewing girls, which, after preliminary neuralgic-like pains, come on at night. These patients relate that they are awakened out of their sleep by the severity of the painful cramps. He has also laid stress upon certain cramps of milkers which originate in an apparently different manner from the most classical milker's cramp. Perhaps they are of the same nature, seen either at a different stage of development or in an individual of different neural tendencies. Cases of atrophy of the thenar, hypothenar, and interossei muscles are known in milkers, and it is certain that peripheral influences cannot be rejected in the interpretation of certain milker's cramps.

By reason of these and other factors it is not possible always to draw sharp lines between the stages in the development of the professional cramp. Each case seems to travel a slightly different path, and one sees the patients all tending in the same general direction, but at different stages and with varying syndromes. The accidental occurrence of pressure neuritides constitutes one of the commonest accessory factors.

Inasmuch as *writer's cramp* is the most frequent of these, it may serve as an example. In the complicated muscular coördination of writing every individual follows his own methods, and no two are ever alike, but, although the actual writing may be different in a million individuals, the general mechanism is largely the same. In general three trends are manifest in writing. There are those who make great

use of the smaller movements of the fingers, those who write largely from the wrist, and those whose movements are still wider, employing the whole arm, freehand, as it were. The general grasp of the pen is about the same with all. Those who write a great deal rapidly, such as newspaper men, usually acquire a very great flexibility in their writing, and use all mechanisms in turn. Thus, they can rest certain groups of muscles, and still carry on the writing movements.

Most observers are in accord in ascribing a greater tendency to the development of writer's cramp in those who make greater use of the finer finger movements. Those who write from the wrist and forearm are less liable, while the large freehand writer is usually immune. Gowers is especially emphatic on the harmfulness of resting the hand on the fingers. While these factors of finger position do play a part, they are not *the* most important, and the large arm writer is not necessarily immune. It is by reason of other factors that the training to large arm movements in a "crammer" is often futile.

Writer's cramp is no recent disease; above all, it is not a creation of the steel pen, as has been assumed by some good observers, nor of any special type of penmanship. That its clearer description should have coincided more or less with the introduction of the steel pen is accidental, for in the early days the disease was described as occurring among those who still adhered to the use of the old-fashioned quill, and it is known among the Chinese, who still use a brush. The mere fact of the widespread occurrence of so many different kinds of occupation neuroses is enough to negative any such type of inference. Nor can full adherence be given to the view that the disorder originates entirely from a faulty manner of holding a pen or of the use of certain muscles in a definite way in writing. The trouble lies farther back, although it is readily conceded that certain faulty methods of writing are more arduous than others, involve more muscular action, and are likely to cause greater fatigue.

Clinicians have described four chief types—the spasmodic, the tremulous, the neuralgic, and the paretic forms—but these signify little, since nearly all cases show more or less of all of these symptoms. Writer's cramp, in its strictest sense, shows the spastic type in its purest forms.

In a well-developed case the patient, after perhaps years of gradually increasing difficulty, after writing for some minutes, or perhaps hours, feels a sense of stiffness in his writing. He has neuralgic pains in certain muscles, at the shoulder-joint, the wrist, or in the thumb. A certain rigidity seems to pervade the strokes, and he notes a marked increase in tension in the thumb which grasps the pen. The index finger becomes unruly, the pen is liable to slip, and firmer grasping is resorted to to hold the pen in place. After a short time of rest he may be able to *résumé* his writing for some time before this grade of difficulty again becomes manifest. With increasing severity the troubles become more marked; the firmer grasping of the pen becomes a spasm, at times one of flexion, but more often an extension (Canstatt's two types), the index finger straightens, the pen becomes more upright, the thumb is strongly contracted to hold the pen against the hand, the spasm extends, the thumb

straightens out, and the pen can no longer be held. The writing becomes more and more irregular, scratchy, and scrawly until it becomes illegible.

Other fingers may be involved; in some cases all the fingers are extended, the wrist flexed, the arm brought down to the side of the body, while severe pains radiate throughout the entire arm. Brusque pronation or supination of the arm may then follow. The whole writing apparatus becomes fixed in a tonic (sometimes clonic) battle between agonist and antagonist, all acting out of concert, and mutually disrupting the harmonious relations of a well-planned coördination. Many patients write for years, with but the sense of stiffness and some pain. Others, on the contrary, develop acute spasms, and no sooner commence to write than the cramp develops. In most of the fully developed cases considerable mental excitement is present. Vascular disturbances may be distinct.

The character of the spasms is usually tonic, and they develop slowly. Some patients have jerky movements somewhat resembling clonic contractions, but these seem less common. Tremor is an almost invariable accompaniment, and there are writers for whom only tremors exist.

It is usually conceded that of the predominant types, the spastic form is most frequent; the feeling of great weakness and soreness in the muscles, which is antecedent to all forms of writer's cramp, is not considered here. The spastic form alone, or in combination with tremors makes up about 40 per cent. of the cases. Berger,<sup>1</sup> in his analysis of 64 cases, found that in 24 there was a pure spastic type; 10 showed the paralytic type, 8 had tremors, while the remainder, 22, had combinations; over 50 per cent. had cramps. In Remak's 42 cases there were 9 cases of true writer's tremor; 32 showed more or less spastic symptoms. He observed only one pure paralytic case. Gowers noted the rarity of paralytic forms.

It is highly significant that the moment the pen is laid aside the cramp ceases, the patient can move his hand or arm in any desired direction, and apart from, at times, a mild sense of tire in the affected muscles, all signs of previous difficulty vanish. The patient can carry on coördinated movements involving the very muscles which were in such a painful state of contraction while performing the single act of writing. Benedikt is stated to be the first observer to record the tendency that exists for the disorder to spread to other frequently repeated intricate coördinations.

In the *tremulous type*, which in its pure state is comparatively rare, the patient's hand begins to shake with the act of writing, the writing is tremulous and shaky, like that of an old person, the whole arm may be involved in the tremor, and patients are known who, without pain or other disturbance, have written such a tremulous hand for thirty or forty years. They have never lost the power of writing, but must always write with a distinct tremor.

In the *paralytic type*, which is rare in the pure form, the patient loses the power to grasp the pen. There is a stage of rapidly advancing fatigue and weakness, up to a point when paresis without spasm or tremor, and often without severe pain, save that of tire, is reached. The fingers become stiff and inactive, as though nailed to the table. After a few

<sup>1</sup> *Beschäftigungsneurosen*, Eulenberg's *Encyclopädie*, 1885, vol. ii.

moments' rest the patient can resume his writing, to be again interrupted by the rapid advance of paresis. The localization of the paretic muscle groups may vary. Berger reports a case of isolated shoulder and upper arm type. Simple abducens pollicis paralysis is known. Duchenne's case of infraspinatus palsy is classical; here the patient could write if the paper was pulled along by the left hand, but the carriage of the arm across the page was impossible.

**Sensory Disturbances.**—Such are usually present if only as a distressed sense of tire, of pressure, of tension. Enough has been said of the neuralgic pains and the pains of tonic contractions. Anesthesia and paresthesia are present, although infrequent; and hyperesthesia with painful nerve trunks may be present, especially in the neuralgic types. Such painful nerve trunks with the occurrence of Lasègue's symptom in the lower extremities may clear up the diagnosis in mild alcoholic cases.

Vascular disturbances, either local, as angiospastic phenomena with cold hands, or as paralytic phenomena with localized sweating, are rare. General anxiety with vascular disturbances is not as infrequent. Electrical changes are not of frequent occasion; when present they indicate the neuritic nature of an occupation palsy, and thus aid in the diagnosis.

**Course and Prognosis.**—The outlook is bad if one has in mind only those cases which are construed to be occupation palsies in the narrow sense. When one takes into consideration, however, all of the forms, especially those of widely varying etiology, the prognosis is good. This is especially true for the alcoholic and pressure types, while in the purely mental types the reverse is true. Most patients with fairly well-developed professional neuroses run an up-and-down course with fairly well-marked chronicity. With improvement in general strength and rest, the professional movements are better carried out, and vice versa. Some patients rest for a year or more, and develop their neurosis within a week after resuming their occupation. In such cases the mental factors are dominant. These patients need psycho-analysis, not rest. In some patients cure seems impossible, either due to defect on the part of the patient to grasp the rationale of a course of therapeutic effort, or a lack of proper analysis on the part of the attending physician. In severe cases of writer's cramp the course is especially obstinate, but not always hopeless.

**Treatment.**—Personal factors enter so largely into this that the individual must always be borne in mind. Occupation neuroses develop on the one hand on a basis of overwork of certain muscular coördinations, without any other complicating factors; at the other extreme, patients will be seen in whom the psychical factors are predominant or have become so, and the fatigue element is practically nil; occupying a partial middle ground are those in whom constant, minute maladjustments are to be borne in mind. Few individuals will present examples of the extremes; the vast majority will present combinations in varying proportions of overwork and mental attitudes with certain intermediary asthenia.

There is little doubt that most patients—save those in whom the psychical features are markedly developed—will receive benefit from the general methods of increasing physical vigor. A sea trip, change, etc., are here indicated. Alcohol and tobacco are to be reduced to a minimum.

Abstinence from writing, in the writer's cramp form, is the first requisite. Many clinicians, Gowers among them, recommend the advisability of teaching their patients to write with the left hand, thus affording the right hand an opportunity to rest. This may effect a comparative cure, but not infrequently such patients develop double writer's cramp. Such an evolution is presumptive evidence of a large psychic element, and for such the methods of Meige are particularly advisable. Methods of reëducation of the writing are also desirable in other cases; changes in the form of pen, using a large penholder, stub pens, changing the slant of the writing, learning to rest one's entire arm on the writing table, and to utilize the larger muscles in the writing act—these, with general hygienic tonic procedures, are at times sufficient to afford sufficient variation for an affected individual to just steer clear of an actual breakdown. Great care must be exercised in this direction. With definite mental features in the case, such schemes are a waste of time, and may be bad psychotherapy.

In the large class in which accessory elements play a part, *i. e.*, congenitally weak muscles, pressure and toxic neuritides, prophylactic measures combined with direct methods of nerve and muscle treatment are needed. Strychnine, calcium salts in the form of hypophosphites, iron, atropine, galvanization, mechanotherapy, massage, etc., are useful. Bier's hyperemic procedures are helpful.

Special electrical methods are futile. Many patients are apparently benefited by the Leduc oscillatory currents. The rôle played by electrical stimulation is difficult to estimate. In those patients for whom such electrical stimulation acts as a general tonic, its action is evident; in others the suggestive feature should not be overlooked; in others it is useless or worse. All the writers who find electricity beneficial report that it must be used for months, which is largely indicative that it is solely a general measure and really has little direct value. Psychoanalysis is of great service for the strictly psychogenic cases, and a great many are such.

### TETANY.<sup>1</sup>

**Definition.**—It cannot be said at the present time just what tetany is, or rather, perhaps, it might better be expressed, that so many different tetanies are known that it is evident that, like epilepsy, it is not a single affection but many; a symptom grouping, very variable as to completeness, which is best regarded as a more or less localized manifestation of a convulsive nature, due to a large number of exciting causes, possibly with some single and unique underlying factor. Whether such a position can be maintained for the so-called idiopathic epidemic form remains to be seen. If the present view be maintained, however, the term tetany must be rejected as a disease conception in the vast majority of the conditions which now go by that name, or perhaps preferably a binominal nomenclature might be adopted which would specify the type under consideration. Thus, it might be advisable to advocate the use of the

<sup>1</sup> Falta, *Die Erkrankungen der Blut Drüsen*, 1913; Phleps, *Die Tetanie*, Lewandowsky, *Handbuch*, 1913; Biedl, *The Internal Secretions*, second edition, 1913.

terms tetany idiopathica, tetany gastro-enterica, tetany thyroidea, tetany toxica, including bacterial and chemical toxins, tetany grvida, etc., until it may be shown just what underlying factor is responsible.

It is highly probable that tetany must be regarded as an example of hyperfunctioning of certain parts of the motor mechanism in which altered muscular excitability is primary, while special neurological features play a secondary rôle, and yet these latter give to the muscular manifestations their special trend. Here particular attention is paid to the etiological factor, since the clinical manifestations alone are not always sufficient to distinguish the form of toxin present.

Frankl-Hochwart,<sup>1</sup> speaking more particularly of the adult forms, is more didactic in his presentation. Tetany, he considers, has as its main symptoms tonic, intermittent, bilateral, often painful cramps, which, without, for the most part, any loss of consciousness, involve the muscles of the upper extremities, particularly the hand, which is held in the obstetrical position. The muscles of the lower extremities may also be involved, those of the larynx, of the face, and of the jaw, seldom those of the chest, abdomen, neck, diaphragm, or tongue. In rare cases the eyeball muscles are implicated, as is also the bladder. In the sensory sphere paresthesia and pains are present, while hyperesthesia occurs now and then. Pressure upon the brachial plexus may give rise to an attack (Trousseau); hyperexcitability to electrical currents is present (Erb); mechanical hyperexcitability of the muscles and motor nerves is observed (Chvostek), while the sensory hyperactivity to mechanical and electrical stimuli is also present (Hoffmann). The psyche is usually uninvolved, mental disturbance being found only now and then. In the chronic and repeating forms secretory and trophic disturbances occur, such as increased perspiration, reddening of the skin, swellings of the joints, mild œdema, falling out of the hair and nails, discoloration of the skin, urticaria, and herpes. Dyspnœa may supervene; polyuria and glycosuria are rare accompanying symptoms. Abortive and incomplete forms this author designates as tetanoid.

For didactic purposes Frankl-Hochwart divides tetany into simple acute forms and chronic recurring forms. A further division of form occurring in children and in adults is made. Tetany of the adult he groups into eight classes: (1) Tetany idiopathica—tetany of otherwise healthy individuals—workman's tetany. This is the form which seems to occur epidemically as an acute, or acutely, recurring affection in certain cities, notably Vienna, Heidelberg, etc., principally in the early spring months, and among certain handworkers—tailors, shoemakers, etc. (2) The tetany of gastric and intestinal affections. (3) The tetanies of acute infectious diseases, typhoid fever, cholera, measles, scarlet fever, etc. (4) The tetanies of acute poisoning, chloroform, morphine, ergot, phosphorus, lead, renal and genital substances. (5) The tetanies of maternity (pregnancy, parturition, and nursing). (6) The tetanies of parathyroid involvement. (7) The tetanies of other nervous diseases, exophthalmic goitre, brain tumors, cysticerci, syringomyelia, etc.

<sup>1</sup> *Die Tetanie der Erwachsenen*, 1907.

Whether all of these have a common etiological factor is a problem that has not yet been definitely solved. It would seem that from the work of MacCallum a deduction might be made allying the various forms—namely, that a disturbance of calcium metabolism of the body is responsible for the hyperactivation of the neuromuscular responses which results in the tetany spasms.

**Incidence.**—Tetany in its different manifestations is undoubtedly rarely seen and is even less frequently reported. In undeveloped phases the tetanoid reaction is comparatively frequent in children. The instances of gastric tetany are probably the most frequent, while the pure epidemic form has not been encountered in the United States. In Griffith's study only 77 cases were found recorded, while Howard's later collection brings the American cases to 154 in 1907. Thus, so far as clinicians in English-speaking countries are concerned, tetany may be considered as being infrequently seen, but even in countries in which it is thought to be epidemic it is rarely observed. It is undoubtedly often overlooked and at times confused with hysteria.

**Etiology.**—At least two factors are concerned in the general hyperexcitability of the neuromuscular apparatus and the neural trend that determines the manifestations of this hyperexcitability in certain groups of muscles and for certain combinations. So far as the first factor is concerned, it would seem that a fairly clear concept has been gained, particularly through recent work, while for the second no reasonable explanation is forthcoming.

As regards the *epidemic tetanies*, Schulz and Hoffmann advocated the thesis that they were due to disturbances in the thyroid gland, this was in the time when the parathyroids were unknown. The fact of the close anatomical relationship of the two glands in man and many carnivora served to hide the deeper truth of the separation of their physiological functions first brought out by Gley in herbivorous animals, in which the glands are anatomically separable and thus capable of exact experimentation. It is unnecessary to go through all the steps which have led to the knowledge that the parathyroids play an important rôle in the functions of the body, and that the peculiar muscular hyperexcitability seen more particularly in tetany is in some manner related to these glands especially in regard to their insufficient action. It is possible that other anomalies of neuromuscular activity are correlated with these glands. Lundborg has advocated wide hypothetical possibilities; even the peculiar motor manifestations of a group of mental disorders, catatonias and their allies, may have some light thrown on them through this avenue. More recent workers have not rested on the apparently certain foundation that tetany is essentially a manifestation of perverted parathyroid activity, but have sought to bring the anomalous forms into conformity with this conception, and, further, to obtain a more fundamental insight into the essential features of the disturbed neuromuscular reaction. Whether the work of MacCallum and Voegtlin<sup>1</sup> has definitely solved this problem is to be determined, but it would appear that the essential

<sup>1</sup> See *Jour. of Exper. Med.*, 1909, vol. xi, for summaries along the lines of chemical research.



factor has been found in the relation of the parathyroid to the calcium metabolism of the body. The hyperexcitability of the neuromuscular apparatus is primarily due to a lack of calcium in the blood, and this is due to a relative or absolute insufficiency of the parathyroid glands. Just how the calcium exchange of the body is controlled by the parathyroids, and what rôle in such control is played by other glands, is not yet known. The conclusions of MacCallum and Voegtlin are so important that they are worthy quoting freely:

"Tetany occurs spontaneously in many forms, and may also be produced by destruction of the parathyroid glands. These glands are independent organs with a definite specific function; whether this function is intimately related to that of other organs of internal secretion is not proven. Failure to produce tetany experimentally is probably due to the fact that some parathyroid tissue remains after an apparently complete extirpation. A small amount of parathyroid tissue is sufficient to prevent the development of tetany.

"In tetany there is apparently some disturbance of the composition of the circulating fluids ordinarily prevented by the secretion of the parathyroids, which disarranges the balance of the mineral constituents of the tissues. Possibly this consists in the appearance of an injurious substance of an acid nature, for such tetanies may be relieved by extensive bleeding, with replacing of the blood by salt solution. No actual poisonous material has, however, been demonstrated by the transference of the blood of a tetanic animal to the veins of a normal one.

"Numerous researches have shown the important relation of the calcium salts to the excitability of the nervous tissues. Their withdrawal leaves the nerve cells in a state of hyperexcitability which can be made to disappear by supplying them with a solution of a calcium salt.

"Tetany may be regarded as an expression of the hyperexcitability of the nerve cells from such cause. The injection of a solution of a salt of calcium into the circulation of an animal in tetany promptly checks all the symptoms and restores the animal to an apparently normal condition.

"Studies on the metabolism in parathyroidectomized animals show: (a) A marked reduction in the calcium content of the tissues of the blood and brain during tetany; (b) an increased output of calcium in the urine and feces on the development of tetany; (c) an increased output of nitrogen in the urine; (d) an increased output of ammonia in the urine, with an increased ammonia ratio in the urine; (e) an increased amount of ammonia in the blood. Much of this affords evidence of the existence of some type of acid intoxication. Its effects are, however, not neutralized by the introduction of alkaline sodium salts, and may perhaps be regarded as especially important in producing a drainage of calcium salts from the tissues which can be remedied by the re-introduction of calcium salts.

"In general, the rôle of the calcium salts in connection with tetany may be conceived as follows: These salts have a moderating influence on the nerve cells. The parathyroid secretion in some way controls the calcium exchange of the body. It may be supposed that in the absence of the parathyroid secretion, substances arise which can combine with

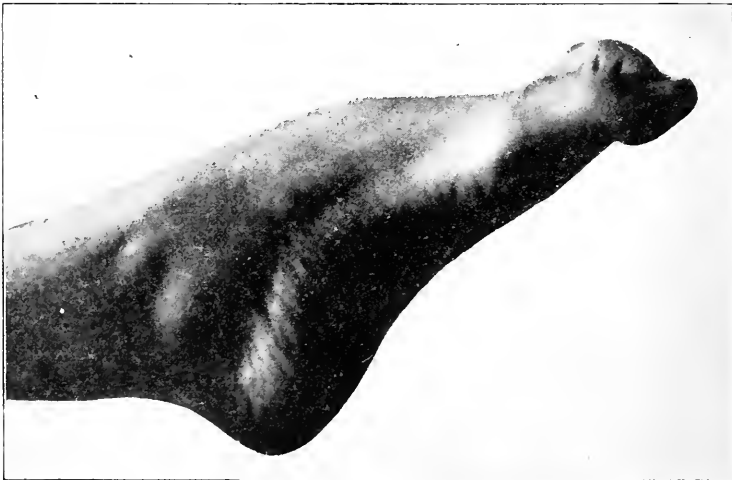
calcium, abstract it from the tissues, and cause its secretion, and that the parathyroid secretion prevents the appearance of such bodies. The mechanism of the parathyroid action is not determined, but the result,

FIG. 41



Tetanic spasm, showing flexion of wrist.

FIG. 42



Tetanic spasm, showing plantar flexion of foot and toes.

the impoverishment of the tissues with respect to calcium, and the consequent development of hyperexcitability of the nerve cells, and tetany is proven. Only the restoration of calcium to the tissues can

prevent this. This explanation is readily applicable to spontaneous forms of tetany in which there is a drain of calcium for physiological purposes, or in which some other condition causes a drain of calcium. In such cases the parathyroids may be relatively insufficient."

It has been shown that salts of barium and of strontium are capable of controlling the spasms of experimental tetany. While this does not constitute a real difficulty in the interpretation of the results just cited, it remains to be shown just what pharmacological relationships these salts have on nerve cells. In view of their close relation to calcium, it may be that they temporarily take on the function of calcium.

The second series of factors still remains to be solved. Why is it, if the facts just quoted bear an essential relation to tetany the disease reaction seems to be strictly localized? Is this simply bad observation on the part of the clinicians, or are there local conditions of diet, water supply, etc., that bring about changes in the intake of calcium? Such conditions are known to prevail for certain infants fed on cow's milk.<sup>1</sup>

Why do certain workers, tailors, shoemakers, etc., suffer and not others? Is Frankl-Hochwart's suggestion regarding the peculiar position adopted by these workers of value in relation to the blood supply of the organ? What have gastric dilatation, gastro-intestinal disorders, etc., to do with the calcium intake? Are there perversions of gastric chemism that favor calcium exclusion? This must be determined in order to explain these cases, and why should certain gravid women develop a parathyroid insufficiency? The observations of Lange, Jeandelize, and others on thyroid activity at this period are only suggestive. Still further, why should the hyperexcitability not be more generalized, and bring about general epileptiform conditions instead of very localized spasms, and further only in certain groups of muscles. These must be more thoroughly explained before the general hypothesis can meet with unqualified recognition, for the symptoms seem to call for a specific rather than a general type of poisoning.

**Pathology.**—Concerning the histological changes the present view excludes a specific pathology. The insufficiency of the parathyroids, be it relative or complete, may be brought about by a great variety of lesions. Such have been described by various observers, and each, in turn, has been considered of specific significance. These in reality offer evidence in favor of the parathyroid insufficiency hypothesis, but go no further. In the minor grades of tetany in children, particularly in so-called spasmophiles which Frankl-Hochwart regards as tetany, the findings of Yanase in Escherich's clinic are illuminating. Here hemorrhages in the parathyroids seemed fairly constant findings, and offer an explanation of the galvanic hyperexcitability. At the other extreme one finds the absolute insufficiency tetanies in experimental parathyreoopriva and in the rarer cases, such as Pool's in man. Here the parathyroid insufficiency is established, and the calcium treatment restorative. In acute epidemic forms thyroid (and probable parathyroid) involvements are known. Tumors, tuberculosis, and a host of other changes in the

<sup>1</sup> See Escherich's Monograph, *Tetanie der Kinder*, 1909, for summaries.

thyroids have been described. It will probably be found that in most of these the parathyroids are likewise implicated. Thus, in exophthalmic goitre a combination of thyroid and parathyroid symptoms is often present. In many tetanies pure thyroid symptoms appear.

The relations of the ganglion cells in the spinal cord which stand in direct connection with the sympathetic system are in need of analysis. Only a few of these groups have been localized, for instance, Jacobson's localization for the Klumpke syndrome.

**Symptoms.**—Considerable variation is to be found, but in general four types of symptoms are observable in the fully developed attack. These are the muscular spasms, which may go on to an exhaustion paralysis, or paresis; the Trousseau phenomenon; increased electrical excitability, or the Erb's symptom; and mechanical hyperexcitability of the muscles—Chvostek's sign. In some patients one or more of these may be missing. Abortive forms, so-called, may present even fewer signs. On the other hand, a richer combination of symptoms, apparently closely related to the general disorder, may be encountered. Sensory disturbances, anomalies of circulation with œdema, of respiration with cyanosis, and of temperature are sometimes found. True psychoses, perhaps indistinguishable from the hysterical confusions, are found. Trophic disorders of the skin, hair, and nails occur. In some rare instances widely confused phenomena resembling epileptic seizures occur.

**Muscle Cramps.**—The most persistent feature consists in a characteristic form of muscular spasm. It occurs chiefly in the extremities, mostly involving the flexors. The spasm is tonic, generally bilateral, and is usually induced by some irritation, mechanical or electrical. Over-exertion, exhaustion, changes in temperature, acute diarrhoea, or emotional excitement may precipitate an attack. Consciousness is involved only in certain forms (parathyroid tetany); pain may or may not be present. The small muscles of the hand are usually first implicated, perhaps after tingling-like prodromata. There is marked adduction in the interossei and the thumb. The hand usually takes a very significant position—that of the "obstetrical hand." It is also described as a "penholding" position. In many mild attacks only the thumb may be involved, and in others the hand alone. Sometimes the hands are closed, making a fist. Flexion at the wrist may follow, the arms then being folded across the chest, or they may be held up in the air or down at the side. Such wider movements occur in the more severe attacks only. A somewhat similar series of flexor cramp-like movements may occur alone or in combination in the lower extremities. Talipes equinovarus, inversion of the foot, and plantar flexion may be present. The legs may be flexed on the thigh, and the thigh on the pelvis in the most severe instances. The contractions in the lower extremities are rarely as severe as in the upper, and frequently are lacking altogether. Eschrich considers the classical carpopedal spasms to be rare in children.

In Frankl-Hochwart's series of 122 cases, only 70 showed cramps in the lower extremities. Occasionally a patient has cramps in the upper and paresthesia in the lower extremities. Frankl-Hochwart makes the assertion that there are no cases on record of tetany confined to the lower

extremities. In Howard's series,<sup>1</sup> however, and these are from the United States, all four extremities were involved in 61 out of 77 cases;

FIG. 43



Method of producing tetanic spasm of hand by stretching the brachial plexus by forcible abduction of the arm. Note "obstetrical" hand.

FIG. 44



Method of producing a tetanic spasm in the feet by stretching the sciatic nerve by forcible flexion of the trunk on the thighs.

<sup>1</sup> *Am. Jour. Med. Sc.*, 1906, cxxxi, 301.

in 14 the arms alone were involved, while in 2 the spasms were confined to the legs, in which latter respect Frankl-Hochwart's claim is negatived. Other spasms may occur. Thus, risus sardonicus, 11 in 122 in Frankl-Hochwart's series, and trismus, in 10 of Howard's, may both be found, although, as a rule, these rarely occur spontaneously, but are invoked by tapping over the facial nerve region in bringing out the Chvostek or Hoffman phenomena. The back muscles were involved in 24 of Howard's series; at times to such an extent as to cause opisthotonos.

Other muscular spasmodic contractions are rare, although it may be said that any muscular contraction may be expected. It is highly probable that sharper delimitation of the tetanoid from the hysterical and epileptoid spasms is needed, and that closer observations will negative the broader statements now usually accepted. Some of these rare contractions occur in the bladder, the rectum, the diaphragm (which has caused asthmatic attacks), and the muscles of the tongue. *Laryngo-spasm* is one of the commonest forms, especially in children, where it has been held as specific for the rachitic. Whether it is to be regarded as a specialized symptom in tetany, or as an individual expression of other conditions, is not at present definitely determined. Spasms of the eye muscles, sufficient to cause diplopia, occur. In 7 of Frankl-Hochwart's series these eye muscle cramps were present. They occur spontaneously. Generalized convulsions are reported in 13 of Howard's cases; 9 of these, however, were in children.

Considerable emphasis may be laid on the bilateral character of these muscular movements. It is extremely rare in the true tetany reaction to find one side alone involved; Frankl-Hochwart reports two cases only.

An incidental feature of some of the hand cramps is the occurrence of an intervention cramp in the same sense as it occurs in Thomsen's disease. The co-existence of the two conditions has been reported.

These muscular cramps persist for a very variable length of time. Not only do they vary in different individuals, but the same patient in different attacks, or at different times in the same attack may show marked variation. In the majority of cases reported the spasms persist for from fifteen minutes to an hour, and two or three hours is not an excessive period. Hoffmann has reported a persistent cramp which lasted for ten days. In fatal cases the contractions pass over into a lethal continuous spasm.

Clonic spasms are rare, but are known as blepharospasm, spasms of the tongue, etc. Postconvulsive paralysis or paresis is an uncommon outcome. Excessive muscular tire, however, is not rare.

**Trousseau's Phenomenon.**—To be able in a free interval to induce an attack, or to increase the attacks in force and frequency during their continuance by pressure upon a nerve trunk, or upon a bloodvessel, is the essential feature discovered by Trousseau. Frankl-Hochwart has almost annihilated the specificity of the conception by stating that pressure anywhere on the body is capable of inducing the attack. This is true in a relative sense only, since sites of election are of primary importance. These are the bicipital sulcus and over the crural artery. The pressure must be distinct, and not fleeting. It may be necessary

to press for at least five minutes. In Howard's American series this phenomenon was present in about 80 per cent. of 45 cases; in Frankl-Hochwart's, in 62 per cent. of 122 cases. The significance of this symptom is unknown. Is it a result of the combined pressure of artery and nerve, as claimed by Trousseau, or is it a sign of anemia, as claimed by Kussmaul and others? Inasmuch as a bilateral spasm is set up by a unilateral anemia, it seems to call for a wider interpretation, and Schlesinger<sup>1</sup> arrived at the hypothesis that it is a reflex.

Ferenczi<sup>2</sup> upholds the anemia etiology, since he has demonstrated that by simply holding the arms high extended over the head this reduction of blood supply will result in spasmodic contractions. If Frankl-Hochwart is correct in his broadening out of Trousseau's concept, since pressure on a bone may bring about the spasm, the anemia hypothesis seems to lack support. Furthermore, his studies on animals would seem to require in explanation a direct nerve stimulus, rather than an alteration in the blood supply. The real difficulty lies behind in the hyperexcitability of the nerve impulse. Trousseau's phenomenon is often well marked in hysterical patients, and in the more classic types of hysteria.

**Electrical Hyperexcitability.**—This is present in (a) motor nerves, (b) sensory nerves, and (c) nerves of special sense.

(a) *Motor Nerves.*—Erb first showed that minimal electrical stimulation (0.5 to 2 milliamperes) brought out the tetany spasm reaction. Later studies of Weiss and Frankl-Hochwart showed the constant hyperexcitability to the galvanic current, while the reactions to the faradic stream were inconstant. A large number of anomalous electrical reactions occur in the motor nerves of these patients. Thomas<sup>3</sup> has described a paradoxical "catelectrotonus tetanus." With cathodal stimulation to the nerve, with currents too weak to be recorded by the galvanometer, one could notice fibrillary contractions in the muscles, which, with the increase in current, became more marked until tetanus developed. This spread to all the muscles supplied by the nerve and ceased on the closure of the current. Anodal stimulation was negative.

Peters<sup>4</sup> has described a "jumping jack" sign brought about by galvanism of the spine, the anode being applied to the sternum, the cathode over the cervical or thoracic vertebrae; minimal currents (3 to 4 milliamperes, even at times 0.5 to 1 milliampere) may cause the arms and legs to jump at each closure of the current. He found that the contractions may be induced on both sides if the cathode and anode are placed directly on the middle line, whereas, if they are placed on one side of the spinous processes the contractions will be limited to that side. In the mild cases the phenomenon is not induced. In such cases the use of an Esmarch bandage will aid in bringing it out. After lumbar puncture it is impossible to bring out the sign.

(b) Hoffmann's researches showed a marked hyperexcitability of the sensory nerves; mild currents causing paresthesia or pain; while (c) Chvostek, Jr., showed that for relatively mild currents distinct auditory

<sup>1</sup> *Neurol. Centralbl.*, 1892, p. 66.

<sup>3</sup> *Johns Hopkins Hosp. Bull.*, 1895, vi.

<sup>4</sup> *Deutsch. Arch. f. klin. Med.*, lxxvii, 69.

<sup>2</sup> *Ibid.*, 1904, p. 294.

sensations were elicited by opening and closing the current. Taste perceptions react in a similar manner, while the optic apparatus does not show any reactions.

**Mechanical Irritability of Nerves and Muscles.**—Chvostek first showed that simple mechanical stimulation of a nerve trunk is sufficient to induce a spasm. Sharp tapping over a nerve trunk, or even over a muscle, is capable of inducing a typical flexor spasm. The favorite site for bringing out Chvostek's sign is over the facial nerve in front of the ear. It is not always present, and in 40 of Howard's patients tested, only 50 per cent. responded positively, but the figures of Chvostek and of Frankl-Hochwart are much more definite. The latter author classes it as one of the most important signs of the tetany reaction, although it is not always present. He further distinguishes three grades in the Chvostek phenomenon. In the more severe reactions (1) a tap in front of the ear will cause contractures in the entire facial innervated musculature. Even light stroking may set free the muscular contractures (Schultze's phenomenon). In the middle grade (2) a stroke of the hammer causes contraction of the nasolabial folds, while in the lightest grades (3) the corner of the mouth alone contracts after percussion in the classical spot. Chvostek's phenomenon has been found in other conditions, and its frequent occurrence in enteroptosis is of particular significance in reviewing the relation of gastric disturbances to the tetany reaction.

Schlesinger's observations of its occurrence in tuberculosis, chlorosis, neurasthenia, hysteria, and severe gastric disturbances are of interest, since Frankl-Hochwart agrees with him largely in his findings, and even goes further and states that he has found the phenomenon in a number of normal individuals. In these, however, it exists in his grades Nos. 2 and 3. He raises the suggestion, however, that possibly many of Schlesinger's patients really were mild or abortive cases of tetany. Its occurrence in myxœdema (Kraepelin), cretinism (Eiselsberg), and related trophic disturbances is recorded.

A sensory hyperexcitability has been described by Hoffmann and confirmed by many. It manifests itself in a manner analogous to that of the Chvostek mechanical excitability of motor nerves. Percussion over a sensory nerve trunk gives rise to painful sensations or to widespread paresthesia in the entire distribution of the nerve percussed.

**Mental Phenomena.**—Although it is considered characteristic that little or no involvement of the mental functions should be present, certain patients with well-developed tetany show mental symptoms. Frankl-Hochwart has called attention to this, and Kraepelin has observed it. Such mental symptoms have varied from the marked hyperexcitability of the chronic tetanoid patient to acute transitory confusion and delirium—quite in the sense of a toxic delirium—to a fairly advanced degree of deterioration in the sense of secondary dementia, as seen in myxœdema, epilepsy, and related types. The cases reported are still too few and imperfectly studied to permit any generalizations. It cannot yet be decided whether the tetany is to be regarded as primary or secondary in many of these cases, and hence the relationship of the psychosis to the tetany or to an accompanying condition is far from being clear.



**Special Senses.**—Modification of *sight* is a rare complication. Changes in the general eye apparatus seen as a whole are of not infrequent occurrence. Thus, ocular palsies, nystagmus, anomalies of light and accommodation reactions are recorded. Retinal changes are occasional, hyperemia and neuritis having been seen. These are to be regarded as largely incidental in the development of the disorder, and not an intrinsic part. The trophic anomalies known to cause cataract are probably more fundamental; they are an impress of the general trophic disturbance which also sets up the modifications in the excitability of the neuromuscular apparatus, setting free the tetany spasms. *Hearing* and *taste* are rarely involved. Buzzing in the ears is to be regarded as a general widespread disorder, and has nothing to do with tetany itself.

**Sensory Phenomena.**—In addition to the purely motor and electrical phenomena, certain sensory changes are often present. These are more fleeting and less constant, as a rule. Pains during the contraction are usually present. They may be severe, or consist of a slight feeling of tension and drawing. Paresthesia is irregularly distributed and extremely common. Such sensations Hoffmann claims may be induced by tapping on the sensory nerves or by electrically stimulating them. Such paresthesia is especially prevalent in the incomplete tetany cases. Anesthesia is occasionally found. In such instances one should bear pseudo-tetany or hysterical tetany in mind.

**Temperature changes** have been noted for many years, but are infrequent, and probably the result of some infectious process, which is a possible primary cause for the metabolic disturbance in the individual case. Respiratory and circulatory changes are to be regarded from a similar standpoint. Involvement of the muscles of respiration naturally cause primary disturbances. The genito-urinary findings have been inconstant. Blood changes are of considerable interest from the experimental side. MacCallum and Voegtlin have shown a marked reduction in the calcium content of the blood in experimental tetany.

**Trophic Phenomena.**—These are very variable, and one suspects that some bear very little relation to the disorder. Certain of them are probably fundamental. Attempts have been made to explain all these changes on the general hypothesis of a vasomotor disturbance. The development of a cataract, which is not uncommon, is thus explained by Peters; but the general disturbance of parathyroid activity is more important. Falling out of the hair, loss of the finger-nails, dental anomalies, irregular sweating, œdema, joint and tendon effusions, cyanosis, redness, various eruptions of an erythematous, scarlatiniform, measly, herpetic, or urticarial nature, are all seen. Many of the skin eruptions occur in the gastric cases. The trophic disturbances in tetany strumipriva (parathyroidectomy) are to be seen from the same standpoint.

**Tendon Reflexes.**—These are known to be modified in a certain number of patients, particularly in the direction of hyperexcitability. Ankle clonus has been recorded.

**Course.**—Clinicians have recognized arbitrarily three groups of cases in adults, and most modern authors are inclined to follow Trousseau in his classical description. In the *benign* form the sensory phenomena,

such as formication or a simple sensation of heat, may precede the spasm. These are confined for the most part to the hands or occasionally to the feet. The contractures may be fleeting, persisting for from five to fifteen minutes, or they may persist for an hour or more. Often the attack terminates by a recurrence of the sensory symptoms. A period of repose lasting for a quarter of an hour to two or three hours supervenes, and the spasms recur. In some instances two or three attacks a day may persist for several months. These mild attacks may recur at yearly intervals, or, as in some of Frankl-Hochwart's cases, several years may elapse, and then they will recur. These benign cases are usually unattended with much pain. Consciousness is not disturbed, and there are usually no sensory, trophic, or temperature disturbances. Again, more *severe* attacks are observed. These occur more often in young adults, the benign forms having been mostly observed among children. Here the contractions are more violent and more painful. The preceding sensory phenomena are usually more pronounced, and with the increase in the severity of the condition other symptoms may be noted. Headache, malaise, and a rise in temperature of  $1^{\circ}$  to  $3^{\circ}$  may be noted. The affected muscles may show signs of congestion, and localized œdema of the hands and feet may be observed. Other muscles than those of the extremities may be involved. The muscles of the abdomen may be frequently contracted or the sternocleidomastoid and the pectorals; strabismus may be noted, either the external or the internal rectus, particularly the latter, being involved. Trousseau first noted the spasms of the pharynx, the larynx, the bladder, and the muscles of respiration, with severe dyspnœa in the event of the last being affected.

These severe attacks are rarer than the benign ones. Frankl-Hochwart has shown that there is a distinct tendency for the well-marked lighter cases in many instances to become graver, and the good prognosis which most writers have given is now seriously doubted by this observer.

In the *grave form* there is no addition of symptoms. The attacks occur with greater and greater frequency and become more and more intense, and the patients die as a direct result.

**Diagnosis.**—The diagnosis of a classical case offers few difficulties. The presence of cramps in the upper extremities, alone or in conjunction with the lower limbs, with the classical obstetrical hand and the additional evidence supplied by the Chvostek, Trousseau, and Erb signs, is usually sufficient to determine a diagnosis. Frankl-Hochwart would separate the different forms, saying that gastric tetany often offers particular difficulties, especially as a group of individuals exists in whom with gastrointestinal disturbances there is a tendency to cramp-like contractures with the typical hand position. He groups them with the hysterias, rather than with the tetanies, however.

*Hysterical pseudo-tetanies* offer perhaps the most difficult diagnostic problems, especially as mixed and combined forms are undoubtedly known. It is not to be forgotten that some authors enlarge the hysterical group and limit the tetany group, and when it is considered how clumsily at times physicians conduct their examinations for the Chvostek, Trousseau, and Erb signs, especially overlooking the opportunities for sugges-

tion in their examinations, it is perfectly understandable why such variances exist. Apart from tactless suggestive influences, however, Curschmann, as well as many another, has shown that the so-called cardinal symptoms are in reality not absolutely trustworthy. It is at times almost impossible to make a diagnosis from pseudo-tetany on a hysterical basis. The mental influence of accident insurance, litigation, simulation, aggravation, etc., should receive careful consideration. Especially is one liable to cast too much discredit upon the patient's reliability if these factors appear. The extensive literature quoted by Frankl-Hochwart should be consulted in this connection.

*Tetany strumipriva*, or, better, *parathyreopriva*, as suggested by Erdheim,<sup>1</sup> offers the most classical manifestations of the disorder, throwing, as well, considerable light upon some of the possible underlying and fundamental features of this peculiar reaction type. Insufficiency of the parathyroids results in convulsive phenomena of the tetany type. Pineles<sup>2</sup> has collected all the cases of tetany parathyreopriva, the study of which is one of the most important of the steps which led to the conception of tetany as a metabolic disorder, in which parathyroid activities are chiefly concerned. Since it is more clearly realized that the parathyroids are embedded in the thyroid tissue, and that the disorders of both may be present, the real difficulties in diagnosis consist in the separation of the symptoms due to disturbances in function of each gland.

**Prognosis.**—The point of view here maintained precludes the possibility of the statement of a general prognosis. Very little is known definitely of the prognosis in infants and children. Most authors agree in giving a fairly good prognosis, although Frankl-Hochwart says that healthy children rarely acquire convulsions, and that the prognosis is not good. In many of these children only one tetany-like spasm has been noted. In others the cramps may persist for weeks and even months. In simple cases the prognosis is much better than in those complicated especially with gastric or intestinal affections. Bronchitis, pneumonia, and occasionally an ascaris infection also determine a less favorable prognosis. Dangerous signs appear with glossal cramps, which may cause death. Recurrences are frequent in those who recover.

Tetany coming on during pregnancy and childbirth usually has a good prognosis. The hyperexcitability of the nervous system may persist for weeks after delivery. In succeeding pregnancies the recurrence of the phenomenon may be looked for. Frankl-Hochwart notes that in succeeding pregnancies the attacks are apt to be milder. It should be borne in mind, however, that sudden death may occur in these cases.

In the cases apparently due to disturbances of the stomach surgical interference has brought about distinct amelioration. Sudden death may occur, and apart from surgical intervention the prognosis is admittedly bad (70 to 80 per cent.). The cases are comparatively rare, however. In severe cases associated with marked gastric dilatation, operation,

<sup>1</sup> *Mitt. a. d. Grenzgebiet. d. Med. u. Chir.*, 1906, vol. xvi.

<sup>2</sup> *Sitz. b. d. Akad. d. Wiss. in Wien*, 1904, 113, Abt. 3, p. 190; and *Mitt. a. d. Grenzgeb. d. Med. u. Chir.*, 1904, xiv, 120. Also consult Pool, *Annals of Surgery*, 1907, xli, for a complete discussion.

if only exploratory, is advisable. The mortality after operation in some dozen or more cases now reported is as low as 30 per cent. The subsequent history of these patients remains to be reported.

Tetany following infectious diseases and acute or chronic poisoning seems to present a favorable prognosis.

The prognosis of tetany thyreopriva depends upon the amount of thyroid gland removed and whether the parathyroids are included. Total extirpation of the entire thyroid tissue and parathyroid is recognized to have a fatal outcome. The entire after-history of thyroid operations, so far as tetany and its prognosis as consequent on the operative procedures are concerned, must be entirely rewritten, since it has been in recent years only that the parathyroids have been reckoned with as an etiological factor. Tetany appears after total removal of the parathyroids, not only in man, but in lower animals. What the outcome of complete parathyroidectomy in man may be with the retention of thyroid tissue is not yet satisfactorily determined. In Pool's case, in which it is possible that the entire parathyroid was removed at the second operation, the patient recovered from the operation and was able to carry on her work for a time. What the ultimate history may be is undecided. We are as yet not in a position to decide as to the definite fate of these parathyroid removals or insufficiencies under calcium or surgical therapy.

The most important class numerically are the so-called epidemic tetanies. Frankl-Hochwart, in his earlier papers, gave a fairly good prognosis in this form, but in later contributions he presents further histories of these patients, and finds that the prognosis is far from being good. Many of them have died, directly or indirectly as a result of their tetany or its causative factor, in from four to eleven years after the onset. Intercurrent disease caused death in many, so that this author only holds that the tetany has perhaps reduced the resistance. Of his many cases, 264 in all, he was able to trace 55 some ten or more years after. Of these, 11 had died early; of the 44, only 9 were healthy; 37 came under personal investigation; 7 had chronic tetany, 19 suffered from tetanoid symptoms; in fact, two-thirds had persistent tetany reactions. This leads him to the conclusion that the prognosis without treatment is in reality very bad, which would be more in consonance with the prevailing point of view that regards the tetany reaction as a result of diminished parathyroid activity.

**Treatment.**—From the standpoint here outlined it may be readily deduced that a general treatment is not a rational mode of approach. The essential feature in the general treatment is the search for the irritant, and this cannot always be successful.

In approaching any specific instance the first question is as to the validity of the phenomenon. This is placed first, because it should be the simplest factor to exclude. If no reason, remote or apparent, can be found which would account for simulation, aggravation, or suggestion, it may be assumed that the excitation of the nervous system has other than a purely psychogenic origin. In rare instances simulation has been a factor; thus, patients have simulated tetanoid contractures in order to obtain the pleasure of chloroform seminarcois.

With a positive diagnosis established, the organ involved should come into review. Inasmuch as parathyroid insufficiency is the most general cause, it is rational to treat those cases, many in children, the whole group of so-called idiopathic tetanies, many tetanies of pregnancy and of thyroid disease, by thyroid and parathyroid preparations. Parathyroid preparations seem to fulfil most of the conditions, yet occasionally the combined thyroid and parathyroid involvement renders the giving of the combined products of more service.

The use of foodstuffs rich in calcium and of calcium salts follows as a natural corollary from the studies enumerated. For the most part, it would seem that such medication might entirely replace the use of the glandular substances themselves. In experimental tetanies the successful effect of the calcium salts has been very striking, and in tetanies in children calcium therapy has given almost uniformly good results. Up to the present time therapeutic experience is not sufficient to definitely prove the durability of calcium medication in the cases in which it seems needed.

The surgical expedient of transplanting parathyroid tissue has proved successful in animal work; its successful application in persistent chronic tetanies in man is clearly foreshadowed by the experimental work on dogs. The technical difficulties do not seem insuperable in view of the ready transplantation of these structures to indifferent parts of the body. Parathyroid transplantation should not be delayed too long. It should be carried out with the onset of severe symptoms and in latent cases that have shown grave signs. In pregnancy, when suspicion of a latent tetany is present, parathyroid transplantation is advisable.

Inasmuch as comparatively definite light has been thrown on the whole subject of the tetany reaction by the researches of MacCallum and Voegtlin in particular, the stomach tetanies, heretofore a particularly obstinate group, offer opportunity for a combined treatment, radical as well as conservative. Useless gastric operations need not be performed, yet at the same time the lines are laid down that point to the time when such operations may be of life-saving service. When obvious gastrointestinal conditions point not only to such irritants in the cycle, it were folly to persist in a line of medication which, notwithstanding its theoretical possibilities, does not give results. Gastrostomy, gastrectomy, gastroduodenostomy, etc., according to the gastric lesion, may be demanded, since the gastric and intestinal mucosa seems to be an important factor in the possibility of calcium intake.

The best methods of administration of calcium salts are matters for individual experience. Foodstuffs rich in calcium, milk, eggs, whey, etc., are clearly useful forms. Injections of calcium chloride, which have been useful in animal work, may be adopted in man. Calcium hypophosphites or other salts to be taken by mouth are available, and intestinal enemata are indicated if other avenues of medication are contra-indicated. The exact dosage remains to be determined.

## CHAPTER XIX.

### PARALYSIS AGITANS. CHOREA. CHOREIFORM AFFECTIONS. INFANTILE CONVULSIONS.

By DANIEL J. MCCARTHY, M.D.

#### PARALYSIS AGITANS.

**Synonyms.**—Shaking palsy; Parkinson's disease.

**Definition.**—Paralysis agitans is a disease of the nervous system characterized by muscular rigidity and muscular weakness, with which symptoms is usually associated a definite kind of tremor, and on which are more or less dependent a deliberation in initiating and a difficulty in carrying out active movements, a characteristic facial expression, a peculiar attitude, and a distinctive gait.

**Etiology.**—**Frequency.**—**Age.**—The disease does not seem to be at all rare in America. According to German statistics<sup>1</sup> there were 37 typical cases among 6000 nervous patients. Although cases of paralysis agitans, most of them doubtful, have been reported before the age of thirty years, and even before twenty years, in the majority of cases the disease does not make its appearance in an individual under forty years of age. Willige<sup>2</sup> found only twelve such cases reported. Gowers found the average of onset to be fifty-three years in males and fifty-one years in females. Cases beginning as late as the seventy-fourth year have been reported.

**Sex.**—The disease is about twice as frequent in men as it is in women (73 males to 42 females, Gowers).

**Heredity.**—The effect of heredity can best be judged by the figures of Gowers and of Berger, who, in 15 per cent. of their respective series, found a history of the disease in more than one member of the family. Five of 19 of Wollenberg's patients gave a family history of nervous or mental disease.

Occupation and station in life seem to have little or no influence in determining the onset, and in the majority of cases the disease does not seem related to any definite determining factor. A direct exciting cause can be traced only in about one-third of all cases (Gowers). The influence of the emotions is certainly an important etiological factor in a group of cases. Gowers emphasizes the importance of fear or fright and reports cases in which such emotion was apparently causative. Anxiety and worry may be mentioned as other predisposing emotional states.

**Trauma.**—The physical influence of trauma, apparent from the attendant psychic concomitants, is well recognized as a determining cause.

<sup>1</sup> *Eulenberg's Real-Encyclopädia*, first edition, Berger.

<sup>2</sup> *Ztsch. f. d. g. Neur. u. Psych.*, iv, H. 4.

Walz's analysis<sup>1</sup> showed in 26 cases, general concussion in 6, wounds (stabs and cuts) in 7, burning and freezing in 1, sprains, twists, and fractures in 4, and contusions in 8. Some connection between the part injured and the locality of the earliest symptoms is almost unexceptional. Trauma may determine the spread of a paralysis agitans already begun. Walz believes that the disease can appear only in a person whose nervous system is deteriorated.

In a group of cases there is a history of overexertion of the muscles, especially of those first affected by the disease. Exposure to cold seems to be a determining cause in another group. The influence of the infectious fevers is probably small.

**Pathology.**—"Chaotic" is the term that adequately expresses our knowledge of the pathology of paralysis agitans. Pathological findings have been manifold, but a definite lesion occurs so inconstantly that at the present time it may be said that the disease has no definite pathology. An exception to this statement seems to exist in the changes in the muscles found by Camp,<sup>2</sup> to whose excellent article the reader is referred.

A peculiar difficulty at the outset of any investigation to determine the pathology lies in the fact that the changes described are not specific, and occur in a variety of conditions. It is difficult, for instance, to differentiate the various overgrowths of connective tissue and neuroglia in the spinal cord from similar changes found in old persons who never have suffered from the disease. Moreover, there is a group of cases which at autopsy shows in the cerebral nervous system no pathological changes whatever. Even the experienced eye of Oppenheim could detect in one case only a "questionable" staining of the column of Goll. Most of the microscopic changes in the brain and spinal cord which have been recorded were reported when the condition was not well recognized, and when the tremor was confused with that of other diseases, especially multiple sclerosis; for instance, one of the cases reported by Parkinson himself was probably a case of multiple sclerosis. Tumors of the brain, sclerosis, cysts, and ependymitis have been reported in connection with the condition, but in these cases either the occurrence of paralysis agitans was a coincidence, or the tremor was not typical. A "symptomatic" paralysis agitans has been written about, but the use of the term "symptomatic" and the reason of occurrence of the tremor have not been made clear. The fact that macroscopic findings occur must be taken into account if we should regard it simply as a symptom complex.

The microscopic changes described affect chiefly the spinal cord; they are many and difficult to separate from those of senility. Many authors agree as to an excess of neuroglial tissue in the cord in these conditions, but differ as to its distribution and its significance. Redlich,<sup>3</sup> for instance, found the chief seat of collections of glial tissue to be in the anterolateral columns, especially around the bloodvessels, while Sanders<sup>4</sup> finds that the neuroglial proliferation is more marked in the gray matter. He emphasizes this as being a different condition than that found in senility.

<sup>1</sup> *Vierteljahrsschrift f. gerichtliche Med.*, 1896, xii, 323.

<sup>2</sup> *Jour. Am. Med. Assn.*, 1909, xlviii, 1230.

<sup>3</sup> *Jahrb. f. Psych.*, xii, 385.

<sup>4</sup> *Monatsschr. f. Psych. und Neurol.*, 1898.

He found not only a comparatively greater increase in the amount of neuroglial tissue in the anterior horn, but also numerous spindle cells in paralysis agitans and cases of senility with a marked tremor.

Dana describes a very moderate increase in the connective tissue within the cord. Tract degenerations have been reported, but they were absent by the Marchi method of staining in all of 14 cases examined by Camp.

Changes in the central canal of the cord, for instance, occlusion and widening, and the presence within it of leukocytes, have been reported; likewise the presence within the spinal cord of amyloid bodies.

Vascular changes reported include arteriosclerosis of the vessels of the cord; miliary hemorrhages into the spinal cord; proliferation of the bloodvessels, which may be surrounded by a zone of leukocytes; miliary aneurisms of the spinal cord; varicosities of the veins in the medulla, with small areas of softening in the oculomotor roots and pyramidal tracts. Numerous authors have emphasized the fact that in the arteriosclerosis found the finer capillaries were more affected than the larger.

Changes have been described in the cells of the spinal cord, in those of the anterior horn, but also those of Clarke's columns. Those in the former include pigmentation, chromatolysis, loss or displacement of the nucleus of the cell, atrophy of the cell, vacuolation of the cell, tumefaction of the nucleolus, etc. Dana describes especially pathological changes in the dendritic process of the anterior horn cells, with consequent diminution in number. This phenomenon was not observed by Camp in any of fourteen cases. Pigmentation of the cells of Clarke's columns has also been described. Lewandowsky has described in the dorsal vagus nucleus pathological changes to account for the disease. Changes have been reported in the Betz cells in the cerebral cortex in a few cases, in connection with changes in the cells of the anterior horns of the spinal cord.

Various degenerations and scleroses have been noted in the peripheral nerves, but these are by no means constant.

In view of the striking phenomenon referred to the muscles in paralysis agitans, it is not strange that a pathological basis of the disease should be sought in the muscles themselves. Certain changes were reported as early as the days of Skoda (1862), who found a fatty degeneration. Numerous observers have since reported atrophy of the muscle fibres, hypertrophy of the same, hyaloid and fatty degeneration and proliferations of the nuclei of the sarcolemma, with smallness of these nuclei. One would expect that some of these might be dependent solely on old age, yet, according to recent observers, the findings in the muscles are distinctive and can be distinguished from changes due to senility.

The findings of Camp, present fairly regularly in a series of nine cases, are quoted: "Many of the muscle fibres are swollen, and in cross-section were round instead of having the normal polygonal shape. There was a marked increase in the number of the nuclei within the fibres, many of which were smaller, denser, and rounder than the normal. These nuclei were frequently in pairs or in long chains. In several places the fibres showed atrophic changes, and in these cases, usually of long-standing disease, there was an overgrowth of the connective tissue. In many changes the fibres had a hyaloid appearance and the longitudinal stria-



tions were unusually well marked, and in many of the fibres there was a tendency to longitudinal cleavage. In one case *trichina spiralis* was found in the muscles, and in this case, in addition to the above described changes, there was an intense interstitial myositis and also a discoidal degeneration of the muscle fibres. Muscle spindles were hard to find in the sections from any of the cases, but in those examined the muscle fibres within them showed similar changes to those surrounding them. The nerve fibres within the spindle were normal, and the intramuscular nerves showed no degeneration when stained with the Weigert hematoxylin stain. These pathological changes were not all of the same intensity in all the muscles nor in different parts of the same muscle." On account of the statement expressed in the last sentence, little stress can be laid on the fact that in some cases of paralysis agitans pieces of muscles have been examined with negative results.

**Ductless Glands.**—The thyroid was formerly the ductless gland most viewed with suspicion as to a possible rôle in the causation, and of late there has been a tendency to hold lesions of the parathyroid gland responsible. It is difficult to separate the changes found in the ductless glands from those due to old age, and from changes due to diseases other than paralysis agitans. Here, again, we are confronted with the fact that in certain cases of paralysis agitans the gland assumed to be the seat of the disease may show nothing pathological. Thus, Alquier<sup>1</sup> examined the thyroid and adrenal glands from two cases of paralysis agitans, and found them negative. Camp, working on an hypothesis of Lundborg and of Berkley, examined parathyroids from two cases and found them pathological, the change consisting essentially in a peculiar infiltration with fat, especially in relation with the bloodvessels. That this is not specific is proved by the observation of R. L. Thomson,<sup>2</sup> who, after a comparison of the parathyroid glands in 9 cases of paralysis agitans with those of 39 cases of individuals dead of other diseases, found that "the parathyroid glands in individuals dying with this disease (paralysis agitans) present no change either in number, size, position, or histological structure that would serve to distinguish them from the parathyroid glands in individuals dying from other diseases." The parathyroid has been the subject during the last three years of many papers, but no more definite result has been arrived at.

Parhon and Goldstein<sup>3</sup> noted a marked preponderance of hematoxylinophilic cells in the hypophysis of a woman who had suffered with Parkinson's disease. Recently, following the work of Kinnier Wilson on lenticular disease, the lenticular nucleus has been carefully studied with positive results in a few cases but with negative findings in most of the cases studied.

**Pathogenesis.**—Many theories have been advanced, but in the absence of a definite anatomical or demonstrated physiological basis, these are little more than speculation. Gowers offers a somewhat hazy explanation by referring the tremor to a derangement of the cortical centres concerned in movements that are naturally brought about by fear. Numerous

<sup>1</sup> *Thèse de Paris*, 1903; *Revue Neurolog.*, 1904, p. 438.

<sup>2</sup> *Jour. Med. Research*, 1906, xv, 399.

<sup>3</sup> *Revue Neurolog.*, 1907, No. 22, p. 1230.

authors hold that the disease is due to an aging of the nervous system before the rest of the body, and point to the fact that the lesions found resemble those of senility. While the theory has some evidence to sustain it, it does not explain the rigidity which is characteristic of the disease or the changes in the muscles. Dana's theory that the dendrites of the anterior horn cells are at fault has some degree of plausibility, although the lesion is not always demonstrable.

The theory that the disease depends on changes in the muscles has much to commend it, and it is possible that the fundamental lesion is some change in the muscle spindles. This latter assumption is rendered the more plausible by the fact that the muscles most severely affected by the tremor are those which contain the greatest abundance of muscle spindles, and that the tremor does not develop in muscles not supplied with the spindles. The subject is, however, rendered complex by our lack of knowledge as to the function of the muscle spindles. Even if we regard them as sensory organs concerned in muscle tone or in muscle sense, it is difficult for us to understand how lesions involving them account for the rigidity and tremor of the muscles, unless we at the same time ascribe some inhibiting influence over these organs by the cells of the anterior horn of the spinal cord. The whole subject is rendered still more complex by the fact that myotonia congenita, a disease characterized by tonic cramp of the muscles on attempt at voluntary movement, presents the pathological changes in the muscles found in paralysis agitans, except that the muscle spindles remain uninvolved. That the disease is an atrophy of the muscles is held by some authors.

**Symptoms.**—As a rule, the disease is ushered in insidiously. There is first a prodromal period characterized by various sensations of heat, weakness, numbness, rheumatoid pains, headache, local oedema, and sensations simulating girdle pains. The symptoms usually affect the part of the body first to be affected by the disease. This period is variable in duration and is sometimes absent, the disease appearing acutely. Palz<sup>1</sup> reports a unique, but not typical case, in which vitiligo was the first symptom.

*Tremor* is the initial symptom in about two-thirds of the cases. At times it is entirely absent (paralysis agitans sine agitatione vel tremore). At first it occurs only intermittently, and usually affects the small muscles of the hand. The fingers, especially the index finger, are alternately flexed and extended at the metacarpophalangeal joints, while the thumb moves to and fro laterally at the same time that there is flexion and extension, the whole resultant movement being somewhat of a circumduction. This movement, in conjunction with the movement of the index finger, produces a "pill-rolling" type of tremor, which is very characteristic. This is usually associated with a flexion and extension of the wrist-joint and a pronation and supination of the forearm. The upper arm is affected less frequently than the forearm. In the legs the calf muscles are most frequently affected. The muscles of the neck may or may not be affected, in which case nodding or rotary movements

<sup>1</sup> *Schmidt's Jahrbuch*, 1905, cclxxxviii, 228.

of the head are produced. The tongue and the muscles of the head and face sometimes share in the tremor; rarely, if ever, the orbicularis palpebrarum. At times the muscles of expression and of mastication are involved. The vocal cords and the muscles of the chest and of the abdomen are affected but very rarely.

The movement varies much as to range, and has a rate of about four and eight-tenths to seven oscillations per second (Gowers); it lessens in frequency as it increases in range; the finer tremor of the early stages is often distinctly quicker than the coarser period of the late period (Gowers). The rate of tremor in the arm and in the leg may vary much.

The tremor continues during rest, is somewhat diminished by voluntary movement, and, as a rule, ceases during sleep and narcosis. It is increased by mental excitement, and may be somewhat controlled by passive movement, or momentarily by the concentration of the patient's attention, as, for instance, when he fixes an object with his eyes. When hemiplegia occurs in an individual with paralysis agitans, as a rule, the tremor ceases in the paralyzed members. This cessation may be transitory.

The one symptom of the disease, which is cardinal, and which, with its associated symptoms, constitutes the disease even in the absence of tremor, is *permanent muscular rigidity*. This rigidity is at once apparent to the examining hand, and is responsible for the attitude and gait of the patient, the delay in executing voluntary movements, the facial expression, and the contractures. It affects especially the muscles of the neck and the vertebral column; but is also seen in the extremities and face. The rigidity usually is marked only when the disease has advanced rather far, but, as Wollenberg remarks, "it may be present subjectively to the patient very early, manifesting itself by a slowness and a difficulty in executing active movements." Associated with the rigidity is a more or less well-marked muscular weakness, which, however, never reaches the stage of total paralysis. This weakness, with or without a sense of stiffness, precedes tremor in about one-fifth of the cases, and occasionally is conspicuous with tremor from the first. This weakness may be manifest subjectively at a very early stage; in fact, it may be considered as one of the prodromal symptoms. Objective weakness, which is manifested, for instance, by the inability of the patient to make much of an impression on the scale of a dynamometer, must not be mistaken for the apparent weakness associated with the slowness and difficulty in executing active movements. The latter seems more or less dependent on muscular rigidity, and may consist in the slowness in the movement itself, or in a delay in the transmission of the voluntary impulse to the muscle. The delay may manifest itself even when the movement is executed with fair rapidity (Gowers).

In addition, in consequence of the muscular rigidity, there is a resistance to passive movement. This becomes greater as the disease advances, and may amount to nearly absolute fixation of a part, as, for instance, the head. This rigidity differs from that seen in hemiplegic and paraplegic contractures, in that it is permanently present and is not increased or called forth by the movement that tests it. Owing to the rigidity, the upper part of the body is bent forward, and there is in consequence a for-

ward, in rare cases a lateral or backward, position of the head on the trunk. J. A. Sicard and L. Alquier<sup>1</sup> report deviations of the spinal column in 12 out of 17 cases of paralysis agitans. These they believe to be due to the muscular rigidity; they may consist of kyphosis, lordosis, scoliosis, or a combination of these deformities. The arms are slightly abducted and the elbows somewhat flexed. The wrists are usually somewhat extended. There may be an ulnar position of the hand resembling that seen in arthritis deformans. The position of the fingers varies; in many cases they are slightly flexed at all joints in the position that they naturally assume during rest; often they are flexed at the metacarpophalangeal joints and extended at the others, from the preponderant contraction in the interossei. There may even be overextension of the last phalanx, most marked in the thumb, perhaps because the tip of the thumb is pressed against the first finger. The rest posture is especially frequent when rigidity preponderates over tremor. When the interossei contract in the tremor the "interosseal posture" is generally very marked. Usually the rigidity can be readily overcome, but in extreme cases the contracture of the interossei may go on to the degree of permanent shortening, so that the metacarpophalangeal joints cannot be passively extended beyond a right angle, just as in contraction of the palmar fascia. Occasionally only one finger (as the index) is thus affected. In the legs the rigidity involves chiefly the hip-joints and knee-joints, causing slight flexion of each and adduction of the thighs. It may extend to the feet and even cause talipes equinovarus and distortion of the toes. Permanent contraction of the muscles is very rare, but the writer has known each foot to be fixed in inversion.

In consequence of the peculiar stooping posture of the patient and the lack of spontaneity in the movements, the term "statuesque" has been applied to the attitude. Tilney<sup>2</sup> describes a characteristic position as follows: (1) Rise of one shoulder and falling of the other; (2) when the patient is sitting the long axis of the head makes an angle with the long axis of the body; (3) one eye is less prominent and the opening between the lids and the pupil smaller on one side than on the other.

The muscles of the face share in the rigidity, and show a lack of expression which has been compared to a mask. The muscles of the eyes are but little if at all affected by the slowness of movement, while nystagmus rarely, if ever, occurs as a symptom. The patient may, however, show a slowness in initiating a movement of the eyes from one point to another. A staring expression in many cases is caused by partial or almost complete absence of winking, due to rigidity of the orbicularis.

The *gait* is characteristic. The patient, if seated, arises with difficulty; the first few steps are slow and shuffling, then the steps become progressively shorter and quicker, until the patient has to come to a stop to prevent his falling forward. This characteristic gait (festination or propulsion) was described by Parkinson. At times the patient, in attempting to walk, makes short steps toward one or the other side (lateropulsion), or he may make the short steps backward instead of

<sup>1</sup> *Nouvelle Iconographie de la Salpêtrière*, 1902, xv, 377.

<sup>2</sup> *Neurographs*, 1911, No. 3.

forward (retropulsion). An individual patient may show both propulsion and retropulsion, which may be demonstrated by gently pushing the patient forward or backward, or by making slight traction on his clothing. The gait was formerly thought to be due to the fact that in consequence of the forward position of the upper part of the patient's body he was "running after his centre of gravity." However, many patients whose heads are turned backward show propulsion, and vice versa. The explanation of the phenomenon is probably to be found in the difficulty in overcoming the rigidity of opposing muscle groups and in the slowness of transmission of controlling impulses to the muscles.

The patient's *voice* is commonly high-pitched and piping, and lacks proper modulation. The patient has difficulty in shaping the first word of what he wants to say, but the succeeding words are enunciated more and more quickly. Müller and also Rosenberg reported cases associated with tremors of the vocal cords. Cisler, quoted by Pelz, describes the vocal cords of 75 per cent. of his patients as approximating the cadaveric position. In very advanced cases the speech is a low "mumble," difficult, if not impossible, to understand.

The tendon *reflexes* may remain normal, or may be increased. Oppenheim and Frank describe a false ankle clonus due to tremor. Oppenheim asserts that a true foot clonus is sometimes present, but that the presence of Babinski's sign is to be regarded as an indication either that the paralysis agitans is symptomatic, or that some other disease exists. The paradoxical phenomenon of Westphal is often present; this consists in a long-continued contraction of a muscle on approximation of its ends; and is seen typically in the contraction of the tibialis anticus muscle on flexion of the foot.

There is a delay in the latent period of muscular contraction, and a lessening of the irritability of the muscles and nerves to the electric current (Borgherini). As a rule, the function of the sphincters is well retained; occasionally incontinence of urine is present. Nystagmus is exceedingly rare. Oppenheim reports a paralysis of convergence of the eyes, which he ascribes to a probable tonic rigidity of both abductors. The same author also reports the presence of von Graefe's sign. Moczutkowsky<sup>1</sup> lays stress on the fact that when the patient's forehead is folded, the return to smoothness is very gradual.

Objective sensory disturbances do not belong to the symptomatology although they may occasionally be present. An exception is the fact that in certain cases the surface temperature of the body may be increased.

Beyond a tendency on the part of some patients to adopt a whining and complaining manner, the mind remains very clear; in fact, the good nature and complaisance of most of the patients, in spite of the severity of the symptoms, is a matter of common observation. Dementia may, however, complicate a case of the disease. Subjective sensations include vague pains, vertigo occasionally, and sensations of heat or cold.

Muscular *atrophy* does not form a part of the clinical picture; a slight degree may be present in long-standing cases. Trophic and vasomotor

<sup>1</sup> Reference in *Neurolog. Centralbl.*, 1897.

disturbances may be present; and the skin of the hand may be smooth, shiny, and swollen. The excretion of sweat may be increased, and salivation and drooling are present. The salivation is probably due, at least in part, to delayed deglutition.

The urine may or may not present an excess of phosphates. The sulphates have been found diminished. The total quantity of urea, as a rule, does not vary much from the normal, but it varies in proportion with the total amount of nitrogen. The urinary toxicity is normal (Vires).

**Forms.**—The modifications of the tremors and distribution of the rigidity have given rise to various clinical types. Hemiplegic, monoplegic, paraplegic, and crossed forms have been described. Charcot writes of a “*forme fruste*,” in which the tremor is very slight and may be entirely absent. Entire absence of the tremor is not such a very rare occurrence. The rate of fibre tremor may very much exceed the usual number of produced movements. The character of the tremor may be altered by the influence of active movements, or it may even be elicited by these movements. Rigidity may be long delayed, in which case the tremor may be difficult to diagnose from that of hysteria or neurasthenia. Hyperextension of the limbs may replace the usual flexion, while a backward position of the head is not unusual. Various diseases (Graves’ disease, myxoedema, tabes dorsalis, hemiplegia) may complicate the disease and make diagnosis difficult. A “*forme rhumatismale*” or “*forme douloureuse*” is spoken of by French authors. At times vasomotor or trophic phenomena (muscular atrophy, oedema, purpura) help to make up the clinical picture. Arterial hypertension has been reported.

**Diagnosis.**—In the advanced stages, as a rule, there is no other affection more readily diagnosed; occasionally, however, a case presents difficulty, as in one reported by Spiller, which had been considered arthritis deformans until a tremor appeared in one of the arms. Other tremors dependent on brain disease simulate those of paralysis agitans so closely that a group of symptomatic shaking palsies has been recognized. At times the tremor resembles that of Graves’ disease, a circumstance all the more puzzling because in certain cases paralysis agitans and exophthalmic goitre co-exist. In this case the former affection may be recognized by the presence of the muscular rigidity and associated symptoms. During the prodromal stages it is difficult or impossible to distinguish paralysis agitans from hysteria or neurasthenia. The mental make-up of the individual should be taken into account, and the possibility of paralysis agitans developing in neurasthenic or hysterical individuals should not be ignored. From the character of the pains in early cases of paralysis agitans, even *tabes* may be considered. The early loss of the tendon reflexes, especially the knee-jerks in the latter affection, together with the other signs of the disease (loss of pupillary reaction to light, etc.), will serve to make the diagnosis clear.

Less easy in some cases is the distinction between paralysis agitans and *multiple sclerosis*. It is especially difficult in those cases of paralysis agitans in which the tremor is increased by active movements. The permanent character of the rigidity in the former affection and the nystagmus, sphincter troubles, scanning speech, etc., of the latter con-

dition will, however, be apparent. *Senile tremor* may simulate that of paralysis agitans closely; it is, however, not accompanied by rigidity, and is more apt to involve the head than is paralysis agitans. It develops, as a rule, in true old age, while the onset of paralysis agitans is in the presenile degenerative period. Oppenheim lays stress on the differentiation of paralysis agitans from a condition of paraplegia senilis dependent on arteriosclerosis of the brain and spinal cord, in which the body is held as in paralysis agitans, but in which true paralysis exists. In some cases the latter condition resembles a pseudobulbar palsy, and the diagnosis becomes difficult, as pseudobulbar palsy may complicate paralysis agitans. Hereditary tremor and toxic tremors can be readily differentiated by the absence of rigidity.

At times it becomes difficult to determine whether a paralysis agitans following an injury is a true condition or simply a neurosis. Each case should be judged on its merits, but collateral symptoms of hysteria speaks against the genuineness of a complicating Parkinson's disease. Oppenheim points out that this neurosis is a persistent affection in itself, but lacks the progressive character of paralysis agitans. Finally, we must distinguish paralysis agitans from the *choreas*, and especially from posthemiplegic choreas, especially as Parkinson's disease may develop on a hemiplegia. The jerky, irregular character of the choreas is absent.

**Prognosis.**—The disease in itself does not seem to shorten life. In certain cases the symptoms tend to progress, both as to extent and intensity, while in others they remain relatively slight.

**Treatment.**—Little or nothing can be done to effect a permanent cure. All efforts should be concentrated to making the life of the patient as comfortable as possible. Suitable diet, bathing, exercise, and rest should be provided. The patient should be kept as quiet as possible, and should avoid all the health resorts likely to cause mental or physical excitement. Electricity should be given as electric baths, especially the dipolar faradic (Oppenheim), otherwise it is of no service.

Friedländer<sup>1</sup> recommends active movements of the extensors, and also passive movements to relax the rigid muscles and to improve the power of their antagonists. This relaxation is well gained by allowing the various members of the body to fall against gravity. At first some difficulty in inducing the patient to relax sufficiently to let a certain part fall may be experienced. After a little experience in letting it fall has been gained it should be raised against slight resistance. The movements should be practised several times during the day, but not for many minutes at a time. This treatment should extend over a long period of time.

Arsenic in the form of Fowler's solution is probably the best agent for maintaining the general tone of the patient.

Hyoscine hydrobromate (gr.  $\frac{1}{200}$  to  $\frac{1}{150}$  three times a day) and duboisine (gr.  $\frac{1}{200}$  to  $\frac{1}{150}$  three times a day), preferably hypodermically, have been recommended for the control of the symptoms, and in certain cases do exert temporary influence. Their use is attended with some danger, as they may give rise to symptoms of intoxication, which is

<sup>1</sup> *Zeit. f. physikalische und diätätische Therapie*, November, 1907, p. 468.

a signal for their temporary withdrawal. Neither of the drugs is a curative agent. The bromides do not seem to be of value.

Berkley recommends extract of parathyroid gland. All preparations of the gland should be kept on ice. "The initial dose of the powdered gland was gr.  $\frac{1}{10}$ , two to four times per day, preferably in capsules; larger doses appear to produce weakness, constipation, nervousness, and even an exaggeration of the symptoms of the disease. The first good effects in the cases treated were noted, as a rule, after fifty to seventy-five capsules had been taken." Berkley<sup>1</sup> now gives parathyroid nucleoproteid, gr.  $\frac{1}{50}$ , in capsules twice daily for at least three months to a year. Roussy and Clunet<sup>2</sup> report good results from radiotherapy of the throat by destruction of the hyperplastic elements of the parathyroid.

### ACUTE CHOREA.

**Synonyms.**—Sydenham's chorea; chorea minor; St. Vitus' dance.

**Definition.**—A disease occurring chiefly in children, due to the effect of an infectious agent, or its toxin, on the central nervous system, characterized by irregular, involuntary muscular contractions, resulting in movements of a purposeless nature, and associated with psychic manifestations.

**Etiology.**—**Age.**—Chorea may develop at any time of life, but it is essentially a disease of childhood and adolescence, and is found most frequently in the later years of childhood. In Osler's analysis of 535 cases, 33 occurred in the first hemidecade, 228 in the second, 212 in the third, and 62 in the fourth.

**Sex.**—The disease is much more frequent in the female sex; according to Gowers the ratio is 3 to 1. In Osler's statistics females were affected in the proportion of 2 to 1.

**Race.**—Mitchell called attention to the absence of chorea in the negro race, and Osler found only 5 out of 175 cases at the Johns Hopkins Hospital. The same author has noted the absence of chorea in the full-blooded Indian. It, however, may occur in the half-breed.

**Climate.**—An attempt has been made by Morris J. Lewis, in the study of cases at the Orthopædic Hospital in Philadelphia, to establish a relationship with climatic conditions (barometric and storm influences), and with apparent success. Other observers, however, working along the same lines, failed in establishing any such relationship. The frequency of attacks prevailing in the spring months can be attributed to other causes. Children are usually of poorer nerve tone and general nutrition after a winter of confinement and school work than at any other period of the year. The strain of school examinations also has an influence in children of a nervous, excitable temperament.

**Emotional Influence.**—One cannot fail to be impressed by the large number of patients with chorea who present a nervous, excitable temperament. There is, indeed, a certain type of child who might be referred

<sup>1</sup> *Med. Record*, xxviii, 1146.

<sup>2</sup> *Arch. d. med. Experimentale et Anat. Path.*, 1910, No. 3.



to as of a choreic temperament. Such children, usually of the female sex, show an extreme condition of motor unrest. All of the movements are quick and extremely jerky. Under excitement the child is so fidgety, the movements of the arms and legs so jerky and apparently purposeless, as to constitute a condition of normal chorea. When such children have Sydenham's chorea, it is extremely difficult to see where the normal movements cease and where the manifestations of the disease begin. This is particularly true during convalescence. On such a soil it takes but little intoxication of the nervous system, from a relatively mild infection, rheumatic or otherwise, to produce a well-developed case of chorea. In nervous children a decided emotion, such as fright, or excessive mental strain, may be the determining factor in the precipitation of an attack. It would be a mistake to consider emotional disturbances as a cause of a clinical syndrome running such a definite course as Sydenham's chorea. In highly emotional children hysterical manifestations or even fully developed major hysteria may complicate the clinical picture of chorea. While the association of the two diseases has nothing in common, there is a possibility of a hysterical child simulating Sydenham's chorea when there has been sufficient opportunity for observation of another case. Epidemics of chorea in schools may be so explained.

**Heredity.**—The influence of a direct similar heredity is very rarely seen. An unstable nervous system, inherited from ancestors suffering from epilepsy, hysteria, insanity, etc., affords a good soil for the development of various functional neuroses. Such an inheritance is, however, not sufficiently frequently met with as to be considered as an essential etiologic factor in the development of chorea.

**Pathogenesis.**—This has been the subject of much discussion, and has led to extensive statistical investigation. Like so many organic and functional nervous diseases of unknown origin, it was for a long time, and is still in many text-books, attributed to fright, rheumatism, exposure to cold and wet, etc. Even the early incidence of the disease does not save it from being attributed to syphilis. As a result of careful study and observation, Sydenham's chorea, as differentiated from habit chorea, organic chorea, and electrical chorea, is now well recognized as a syndrome, the result of the actions of an infectious toxic agent upon the central nervous system. Wollenberg designated the disease as infectious chorea as differentiated from degenerative chorea and other choreiform conditions. It must, however, be admitted that a satisfactory conclusion has not yet been reached concerning the specific infecting agent or toxin. Nearly all the investigations so far recorded are either controversial or disputatious in character, and for that reason lack scientific value. In nearly all of these studies, both clinical and pathological, the authors have assumed a position (necessarily one of theory), and have then written or investigated in support of their position.

The evidence is far from conclusive as establishing a distinct causal relationship between rheumatic fever and chorea. From a clinical standpoint, the statistics vary between 1.6 and 85.5 per cent. In the articles, however, in which the subject has been studied from an unbiassed standpoint, in which there is every evidence of careful investigation and

record by competent observers, there is a fair uniformity in the statistics. Thayer, Osler, Wollenberg, etc., give statistics varying from 20 to 25 per cent., and this even with a wide latitude as to what constitutes rheumatic fever. These statistics, as presented by Thayer, are of especial interest on account of the relationship between rheumatic fever and organic heart disease. Thus, 21.6 per cent. of these cases show an association with rheumatic fever, and 25.4 per cent. show evidence of organic heart disease.

When an analysis is made with the autopsy records, another difficulty presents itself. Uncomplicated chorea is looked upon as having a very favorable progress. The mortality as given by the British collective committee is 2 per cent. Sinkler found sixty-four deaths in Philadelphia in seventy-four years. Taking into consideration, therefore, the frequency of the disease, it necessarily follows that the mortality is very low. The mortality of chorea gravidarum is given as only 20 to 25 per cent., and this is a very rare manifestation of the disease. Admitting therefore, the complication of rheumatic fever in 25 per cent. of cases, it would not be at all surprising if an analysis of the deaths should show a large percentage of cases in which this complication was present.

Other septic processes, which may have endocarditis as a complication, may also find a place in these statistics. When, therefore, Poynton and Holmes take the position that chorea, to use the expression of Duckworth, is a "cerebral rheumatism," and support this position by finding a micro-organism in six cases, this position can only be considered as scientifically correct for a single group of cases. In at least one of these cases, however, the colon bacillus was found in the blood at autopsy. Little stress was laid upon this fact, and he used this case as an argument in favor of his position. A more reasonable conclusion from the evidence of these six cases would be that when a severe type of chorea complicates a rheumatic infection, it is very likely to prove fatal. Various statistics suggest that we are justified in considering the close association of rheumatic fever and chorea in the maximum of 25 per cent. of cases.

It should be remembered that in chorea, like epilepsy, the disturbance of the function of the cerebral motor apparatus may occur as the result of various causes. It would be unscientific to consider that large group of cases in which convulsions begin in children of unstable motor mechanism, and which are the result of any infection, as cases of measles and scarlet fever, because these happen to be the most frequent of the infections of childhood. It seems equally unscientific to consider that chorea or similar derangements of the motor mechanism symptomatic of an acute infectious toxic process are in all cases due to rheumatic fever, because a rheumatic infection occurs relatively frequently as a late infection in childhood. Macalister,<sup>1</sup> using Ross' method for the study of the life of the leukocyte, has shown that while the toxin in the blood plasma of chorea is toxic to the leukocytes of healthy persons, the blood plasma in cases of rheumatism is scarcely at all toxic, and that the plasma from chorea cases was toxic to the leukocytes of rheumatic cases; from

<sup>1</sup> *Proceedings of Royal Society of Medicine*, July, 1909.

which it would appear that the poisons in the two diseases are dissimilar. The writer is fully aware of the slight or negative articular manifestations of rheumatic fever in childhood, but does not agree with some of the observers that every case of endocarditis, pericarditis, and myocarditis in childhood, in the absence of joint manifestations, is to be considered rheumatic. We must consider, on the one hand, that an infection in childhood is likely to be associated with vague pains, and that, on the other hand, gonorrhœa, beri-beri, syphilis, influenza, tuberculosis, erysipelas, pneumonia, and meningitis are associated with arthritic manifestations which are described as "rheumatism," etc.

**Other Infectious Diseases.**—Measles, scarlet fever, diphtheria, typhoid fever, whooping cough, pneumonia, cholera, and smallpox have occasionally been reported as causative factors of chorea. Syphilis, and septic infections, followed by chorea, are also on record. None of these diseases, however, with the possible exception of scarlet fever, occurs with any degree of frequency. The rarity of chorea as a complication of this condition is seen from the statistics of Carslaw, who found only 3 in 533 cases of scarlet fever. The statistics of Osler are somewhat misleading unless properly interpreted. Of the 554 cases reported, the history of scarlet fever was present in 141 cases, without, however, any causal relationship between the two diseases. In none of his cases did the chorea follow immediately upon the scarlet fever. In this connection it should be noted that any condition, whether of an infectious nature or not, if it lowers the vitality and thereby lessens the nerve tone, renders the child more susceptible not only to chorea, but to other functional nervous disturbances.

**Pregnancy.**—This is an important factor in the production of chorea, more particularly in the chorea of adult life. The statistics recorded show the incidence of chorea to be relatively slight after the sixteenth year. Only 16 per cent. of the cases, according to the collective investigation committee's report, occurred between the ages of fifteen and twenty. When, however, chorea occurs during adult life, pregnancy must be considered one of the most important, if not the most important, factor in its production. Chorea may develop at any time during pregnancy, but is most commonly seen in the earlier months. It usually occurs with the first pregnancy, but may not appear until the second. After its development, however, with the first pregnancy, it may, as in a case reported by Féré, occur with each successive pregnancy for as many as five times.

**Reflex Causes.**—Reflex irritation due to various causes, such as intestinal parasites, intestinal indigestion, phimosis, adenoids, and eye-strain, has been given as a cause of chorea. There is no question that irregular movements simulating true chorea are sometimes produced by such causes in nervous children. It would, however, be a mistake to consider irregular and bizarre movements as identical with a true chorea. A study by de Schweinitz has shown a condition of hypermetropic astigmatism in 77 per cent. of choreic children, and he concludes that a refraction error may be a determining cause of a choreic attack in a susceptible child.

**Pathology.**—Lesions either gross or microscopic have not been found in a sufficient number of cases to constitute a definite pathological basis. Minor vascular and perivascular conditions have been described in a small number of cases, and have constituted the basis for the embolic theory. Terminal emboli have been found with more frequency in the central artery of the retina than in the cerebral capillaries; Thomas collected 7 cases. It is not surprising, in view of the frequency of chorea and the large number of cases showing an endocarditis as a complication, that emboli should occasionally be found. The absence of extensive capillary emboli in the cerebral tissues and a more careful study of the bacterial infections, have led most observers to a position adverse to the embolic theory. The other vascular changes, microscopic perivascular hemorrhages, perivascular leukocytic infiltration, hyaline changes of the vessel walls, hyaline spherical bodies, have occasionally been noted in isolated cases. Inasmuch, however, as these same changes are more frequently noted in other conditions without choreic manifestations, they cannot be regarded as constituting a basic pathology.

**Bacteriology.**—The reaction of the tissues of the newborn child to bacterial invasion has been found to be somewhat different from that of the fully developed individual. The central nervous system is at birth in process of development. Relatively few of the fibre tracts have reached their full myelinization. The functions of the nervous system are also relatively incomplete. In its reaction to varying stimuli, the nervous system cannot be regarded as complete in its development and of a stable nature until after puberty. An infectious agent or a toxin will give entirely different reactions at birth, during childhood, and in adult life. In children with a finely balanced nervous system and with a sensitive reaction to infectious agents or their toxins, a condition of disturbed function of the nervous system is more easily produced than in the more rugged type. Poynton and Paine considered the diplococcus found by them in rheumatic fever as the causative agent in chorea. Previous investigations have shown other organisms. In view of these findings we are forced to conclude that the exact bacterial cause of the disease has not been conclusively proved.

**Symptoms.**—Cases of chorea divide themselves naturally into three groups according to the severity of the infection: (1) Mild chorea; (2) severe chorea; and (3) malignant chorea, or chorea insaniens.

**Mild Chorea.**—A prodromal period of a week or more is usually presented. The child is listless, somewhat depressed, nervous, with some anorexia and sometimes anemia. The nervous irritability becomes more marked, and with it slight, irregular, purposeless, jerky movements of one or both upper extremities. This is associated with a pseudo-loss of power in the extremities affected. The pseudo-loss of power, which is dependent on lack of control, is often noticed by parents as the first symptom. A careful testing of muscular power will show usually a normal condition. The tendency to drop articles which the patient may be carrying is not due to a distinct paralytic condition, but to a relaxation of the grasp simultaneously with the development of the choreic jerk. When one arm alone is affected, the disease may progress to the lower

extremities on the same side. In some cases the opposite side does not become affected, and a condition of hemichorea is presented. Usually, however, the opposite side becomes affected, following which the head and face share in the movements. Irregular bizarre forms are sometimes presented in which, for example, the arm on one side and the leg on the opposite side may be involved. Speech is usually not markedly disturbed in the mild form, although those accustomed to the conversation in the child will often notice some slight change—a slowness of the speech, with some hesitancy and irregularity. The movements in the mild form of the disease cease during sleep. An actual loss of power may develop in mild cases. The mentality is also affected, even in mild cases.

*Severe Chorea.*—This form of chorea, which may develop in all its intensity from the onset, or may, on the other hand, be of a gradual development from the mild form, presents a condition of constant, irregular movements of practically all the voluntary muscles. The mild form is often disregarded by parents and not considered as a serious condition, but in this form the severity of the movements and the inability of the patients to help themselves immediately demand attention. Not only are the movements more constant, but they are increased in severity and very often persist to a minor degree during sleep, and in some instances actually awaken the patient. The speech is affected in almost all cases. This varies from a hesitancy and explosive type of speech to a mumbling, and often inability to speak at all. While choreic movements of the tongue are sometimes observed, this symptom is, in the writer's experience, more frequently seen in hysterical and habit chorea than in Sydenham's chorea. Disturbance of speech is often due to irregularity of the respiratory rhythm. When the respiration becomes jerky and irregular it is not uncommon to find irregularity of the heart action with it. Motor weakness is the rule in this form, but whether it is a real loss of power or an interference with purposive action by the choreic movements is often difficult to determine. The mental symptoms are much more accentuated, the child lacks power of concentration and attention, is often irascible, and presents some failure of memory. In this form a slight elevation of temperature may be noted varying from  $0.5^{\circ}$  to  $1^{\circ}$ . A decided elevation of temperature should always suggest the presence of some complication.

*Malignant Chorea, or Chorea Insaniens.*—This may be the terminal complication of a severe form of chorea, or may develop as a distinct type from the beginning. It occurs more frequently as we approach adult life. There is often some source of intense worry or anxiety as a complicating factor in the etiology. In some cases it is a manifestation of an infectious process on a poorly balanced mentality. Pregnancy is an important etiological factor. The motor manifestations become intense, universal, and constant, may interfere with the sleep of the patient, and rapid exhaustion occurs. A confused delirium, with hallucinations, delusions, and maniacal outbreaks, ensues; the temperature rises as high as  $104^{\circ}$ , and a fatal termination is the usual result. This condition is not to be confused with simple chorea or mild types of chorea complicated by simple delusional states.

**Psychic Manifestations.**—Burr states that “although there are no sharp lines dividing the cases, they may be separated, so far as the mental symptoms are concerned, into the following groups: First (and this includes the vast majority), patients in whom there is peevishness, fretfulness, some loss of the power of fixing the attention, and a slight loss of the moral sense shown by disobedience and selfishness. Second, those showing, in addition to the above symptoms, night terrors, and transitory, visual, auditory, or other hallucinations. Third, those with distinct delirium, wild or mild, accompanied with fever. Fourth (and this group is very small when we remember how common chorea is), those showing stupor, or rather stupidity, and an acute dementia, which may follow the condition described under three, or appear without any preceding mental symptoms at all severe, and which is usually accompanied with trouble on articulation not caused by choreic movements of the lips and tongue, but the result of mental hebetude. Fever is usually present for a time at least. Patients of the first and second groups almost always recover mentally and physically; those of the third group frequently die, and those of the fourth usually either die or, recovering from the chorea, remain demented.”

This states the subject of the mental condition in chorea, practically in its entirety. While the maniacal condition has been considered by some to be in the nature of a “cerebral rheumatism,” it can be more accurately stated that when this condition is associated with a high terminal fever, it represents the effect of the infection producing the febrile condition. States of mental alienation with excitement, as indicative of a grave form of chorea with an unfavorable prognosis, should be differentiated from hysteria complicating chorea. Minor hysterical stigmata, as might be expected from the neurotic nature of a large number of these children, are sometimes met with. Areas of anesthesia, hyperesthesia, points of acute tenderness along the nerves and vertebræ, and suggestive pains in the extremities have been described. Triboulet suggests a correlation of the symptoms to the metameric zones of Head. Rarely, hysterical contractures and convulsions, with mental excitement, may occur. The prognosis in these cases, however, is not altered by this complication.

**Paralytic Phenomena.**—A pseudo-loss of power due to the intensity of the choreic movements has been described above. In addition to this, an actual loss of power, which approaches but never arrives at a complete paralysis, has been noted by various observers since Sydenham. West gave to this condition the name “chorea mollis.” The loss of power may precede the development of the choreic movements, may be coincidental with them, may remain after the choreic movements have to a large extent subsided, and may in some cases inhibit the choreic movements to such an extent as to appear to supplant them. The motor weakness may be partial in nature, a monoplegia, hemiplegia, paraplegia, or in very rare instances may be general throughout the voluntary muscles. While this condition is usually associated with very severe forms of chorea, it may, on the other hand, complicate very mild cases. Apart from the paralytic phenomena, the etiology, symptomatology, and progression remain the same as in the usual type of Sydenham’s chorea.

Rondeau has noted muscular atrophy as a rare complication. This exists in cases in which the chorea complicates, or is associated with, a rheumatic affection. He considers it as a reflex joint atrophy. It disappears with the cure of the chorea and the rheumatic fever.

**Reflexes.**—Much conflicting testimony has been presented as to the nature of the reflex activity in this disease. The writer's experience has shown the reflexes to be uniformly active and often quickened when properly elicited. The child, however, must be relaxed, and its attention thoroughly diverted, in order to determine the exact nature of the reflexes. In chorea mollis, when the loss of power is marked, the reflexes may be diminished in the affected part. Triboulet notes a delayed reflex activity. Other irregularities of reflex action are sometimes noted. The more constant of these is a delayed relaxation of the part to normal after the motor explosion (Gordon). The writer has never seen complete abolition of the reflexes as described by Joffroy. This author described the reflexes as diminished, as a rule, and the normal or excited reflex as an exception. Oddo also notes, in a study of 147 choreics, normal reflex activity on both sides in 8, and on one side in 28 cases, and arrives at the conclusion that normal reflex activity only occurs in mild cases, and that usually the reflexes are diminished or suppressed (116 out of 147 cases). He also notes that an exaggeration and suppression of the reflexes may occur at different times in the same case.

**Urine.**—An excess of urine and uric acid has been found in proportion to the severity of the muscular activity. Garrod found urohematoporphyrin in 14 out of 20 cases. Herter reported the presence of hematomorphyrin in the urine not only of chorea, but likewise rheumatism. Glycosuria has also been noted.

**Complications.**—The most important complication, which should not only be kept in mind but carefully sought for during the course of the disease, is endocarditis. The most comprehensive statement of the relationship will be found in a study by Thayer, who found that 25.4 per cent. of his cases showed evidence of organic heart disease. Osler found evidence of organic heart disease in 72 out of 140 patients examined more than two years after the attack; and in a collection of 73 autopsies, endocarditis in 62. Other complications of a rheumatic nature are erythema nodosum, subcutaneous nodules, purpura, etc. Herpes zoster occasionally occurs, and has been attributed to the use of arsenic.

**Course and Prognosis.**—The usual case of chorea runs a course of from six to ten weeks. A mild case sometimes recovers in a shorter period. Not infrequently the course is prolonged for three or four months. Choreic movements may persist for many months—a condition termed residual chorea by Guthrie. This can be explained to a certain extent by the nervous condition of the child, the movements remaining as a habit chorea, and in rare cases may be due to the presence of some persistent peripheral irritation. Recurrent attacks of chorea are so frequent that the physician must be on the watch for them and take measures to prevent them. A boy recently under observation has been affected with choreic movements in the last five years for a longer period of time than he has been without them. The attacks have occurred each spring and fall, lasting three or four months.

The outlook, as a rule, is favorable. Under proper treatment the patients may be assured of a complete recovery, but the parents should be told of the possibility of cardiac complications and the tendency of the disease to return during a period of several years.

**Treatment.**—Chorea at all times, even the milder forms, is sufficiently serious to demand careful attention and supervision. All patients do better, the course of the disease is shortened, and the danger of complications lessened by confining the child to bed during the period of active symptoms. Anything that tends to produce mental excitement should be rigidly excluded. A nurse trained to handle nervous children is a very helpful adjunct to the treatment. The diet should be simple and nutritious, with tea and coffee excluded. In the severe cases isolation is necessary. Care should always be used in permitting visitors; strangers should be excluded from the sick room, and even members of the family when their visits or presence produce undue excitement. School duties and intellectual efforts should be avoided. Gentle massage, with bathing and a warm or cold wet pack, often have a quieting influence when properly administered. It is quite necessary in the poorly nourished, in whom overfeeding is necessary, to keep the muscles in good condition by routine massage, followed during convalescence by passive and restricted movements. These, however, should be carefully watched, and if any tendency to accentuation in the motor phenomena is manifested, they should be decreased or stopped altogether. Electricity is sometimes of value both as a body stimulant and in keeping the muscles in good condition, but often produces too much excitement to be used. Galvanism should be applied in preference to faradism.

The medicinal treatment is confined to alterative tonics and nerve sedatives. In the former, arsenic, in the form of Fowler's solution, is of distinct value. It is questionable, however, whether its value is more than that of an alterative tonic. There is a possibility of doing harm if the administration of the drug is not carefully watched and the cases for its use properly selected. It is a mistake to give the drug in large doses, and it should never be given until the physiological limit is reached. Its full value may be obtained by the administration of small doses; two to three drops gradually increased until ten drops three times a day are taken. It ought never to be administered except when the patient is under the personal observation of a physician. Indiscriminate and persistent use of the drug may result in arsenical neuritis and serious disturbance of metabolism. Donovan's solution may be administered in doses of two to five drops, in older children, three times daily. Strychnine, cimicifuga, and belladonna are sometimes used.

*Sedative* drugs have a decided value in the more severe and active cases. Chloral is the most valuable of these remedies, and is usually combined with one of the bromides in doses of 5 grains each, three or four times a day. In some cases it may be necessary to increase the bromides slowly, to as high as 15 grains, in older children. In other severe cases hyoscine may be used, either by mouth or hypodermically. Antipyrine, extract of cannabis indica, and chloretone have been found to be of value. Chloretone, like hyoscine, has a decidedly quieting influence in some



cases, but the administration of both drugs must be carefully guarded. Such remedies as aspirin, the salicylates, and alkalies are of value in the control of rheumatic symptoms, but have relatively little influence on the course of the disease. Potts found apomorphine to be of distinct value in controlling the symptoms of very severe cases.

Salvarsan by intravenous, intramuscular and intraspinal injection has given uniformly good results, more particularly in severe and resistant cases. This has led Milian<sup>1</sup> to the conclusion that chorea is in all cases due to syphilis.

Lumbar puncture,<sup>2</sup> the subcutaneous injection of cholesterin,<sup>3</sup> thyroidin,<sup>4</sup> and trional have been reported as giving successful results.

In chronic chorea, when the movements persist after the general condition is improved, and in some of the milder forms, treatment by means of a combination of suggestion, at first passive movements and later by means of exercises, may be of distinct value. This treatment has been systematized by Guthrie as follows: 1. Suggestions that the child should lie quietly should be repeated in the soothing manner used by the hypnotist. At the same time the flourishes and wriggles should be gently restrained, and the child will soon learn to lie completely relaxed and flaccid under observation.

2. Passive movements combined with suggestion. It will be found that choreic movements occur directly the child's limbs are manipulated. To correct this condition the patient's hand should be placed between the observer's hands, and raised and moved in various directions, suggestion being made meanwhile that the child should keep his own hand quite still. At first it will be snatched away and flourished as usual, but soon the patient learns to control the involuntary movement by an effort of will which makes the whole limb rigid. This is an indication of improvement, although by no means of a cure; but by degrees the child discovers that the efforts to control involuntary movements need not be so strenuous whilst his limbs are guided and restrained, and accordingly the rigidity becomes relaxed. Until the limbs can be passively moved freely in all directions without exciting either rigidity or spasm no other than passive exercises should be employed.

3. Voluntary movements under guidance. The patient is then directed to perform the same movements as have been passively executed while his hand is still controlled, guided, and helped by the observer.

4. When this can be done without exciting spasms or rigidity, but not before, voluntary movements without control, in imitation of the observer's should be practised.

The movements should be of the simplest character at first, and gradually made more elaborate. They should be executed slowly and steadily, without jerks and flourishes. The chief difficulty is, at this stage, incoördination rather than spasm.

<sup>1</sup> *Bull. et Mem. de l. Soc. des Hôpît. de Paris*, 1912, No. 23.

<sup>2</sup> Richardson, *Bull. de l. Soc. de Pédiatrie de Paris*, 1911, p. 111.

<sup>3</sup> Margarot, *Montpel. Med.*, 1912, xxxv, 621-627.

<sup>4</sup> Roden, *Lancet*, 1911, ii.

Elaborate apparatus is unnecessary. Simply bringing the fingers together from a distance and touching various parts of the body with them are admirable exercises. Precision of movement can be gained and the child kept amused by various toys and games. Colored balls hung by strings on a frame or strung on wires can be used. Then solid squares or cubes can be built in various shapes and forms. Such games as draughts, dominoes, or solitaire can be used for older children. A child may usually be considered cured when he can build a two-story house of cards.

The aim of exercises is to encourage freedom as well as precision in movement. For this reason it is inadvisable to allow the patient to write or to thread needles or to sew. All these require much mental concentration, fixation, and tension of the various muscles employed. There is no objection to freehand drawing on a slate or blackboard, and paper plaiting or weaving may be encouraged. It is important that all exercises should be carried out under supervision, otherwise the child becomes careless and ceases to take any trouble, provided that he can amuse himself, and if neglected often drifts into what may be called "residual chorea." The treatment in this event is by a course of drilling, marching, wheeling, and standing at attention, at word of command in particular, for the child's chief faults are inattention and carelessness, which have to be overcome. Ordinary calisthenic exercises are also useful in these cases. The important feature of "residual chorea" is that the movements are unconsciously performed, and can be immediately controlled by effort of will.

The treatment of the lower extremities is on the same principles as that of the upper. The patient should not be allowed to stand, or try to walk until all movements of the legs can be executed with fair precision while lying down. Such ataxia as still remains should be treated by making the child stand supported by the back of a chair while he places each foot separately in various positions. It is necessary finally to observe that exercises of any description are inadvisable in all acute and recent cases of sthenic chorea. They are only of use when heart, pulse, temperature, appetite, and digestion are normal, all pains and aches have disappeared, and, although general health has seemed fully restored for some weeks, choreic movements still continue.

The principles in this are: (1) That suggestion is of service where there is lack of inhibitory control over choreic movements. (2) Inhibition when acquired is often exaggerated, and has to be regulated by assistance before voluntary movements can be executed. (3) Inco-ordination has to be treated by exercises carefully graduated in the order of their difficulty of accomplishment.

### **HABIT CHOREA, HABIT SPASM, CONVULSIVE TIC.**

**Definition.**—A tic is a sudden, quick, involuntary coördinated muscular action, usually of a purposive type, and partly or entirely under the control of the patient.

Much time and thought have been devoted to the elucidation of the subject of tics. Sinkler, in using the term habit chorea, placed them in the larger group of the choreas. Objection has been made to this on the ground that, being purposive and intentional, they are of an entirely different nature from the movements of Sydenham's chorea. The term habit spasm is equally objectionable, because a large number of them are not in the strict sense of the term of a habit nature; *i. e.*, they do not develop by repetition from a volitional act. While Guthrie separates these two conditions, it is often difficult in the fully developed complex tic to decide whether it began in the form of a habit spasm or not. All investigators agree in separating from the group of what might be termed harmless tics those cases presenting a similar clinical picture, but developing on a foundation of a serious neurosis. In Tourette's disease, which represents the most marked type of this latter group, all the voluntary muscles may be affected. The cases in this group are classed as incurable. A study of a large number of cases of tic will show a neurotic basis not only for the psychic tics (Tourette's disease), but in the simpler forms.

It is likewise true that in many of the simple habit spasms and tics a psychic element plays an important rôle. Patrick has called attention to the fact that the tic movement represents a distinct psychic want—*i. e.*, a sense of relief as opposed to a very decided sense of psychic distress when the movement is inhibited. Each case of tic represents in its etiology and its essential nature some definite disturbance of the nervous balance. As such, each case needs special and individual study.

**Etiology.**—An inherited or acquired neurotic temperament is the important etiological factor in most cases. In children of a nervous temperament, self-control is an essential factor in maintaining a nervous balance and proper functioning of the nervous system. Habit spasms and tics in early life and other functional neuroses in adolescence and adult life are frequently the result of a lack of discipline in the training of a child. The more vicious the heredity, the more need there is for careful guarding of the education and the self-control of the child.

Anything which reduces the nerve tone of a child may determine the development of a tic. In some children tic may develop as a result of a loss of weight, vigor, and nerve tone, below a certain point. In these children such manifestations may be looked upon as an indication for medical supervision and treatment. In finely balanced children a nerve drain from some irritation, such as carious teeth, adenoids, phimosis, intestinal parasites, etc., may be the determining factor. These causes are usually referred to as reflex. Excessive worry, emotional excitement, or other forms of mental overstrain may in some cases be the cause.

**Symptoms.**—The movements differ from those of Sydenham's chorea in their evident purposive character and their localization to a single muscle or group of muscles. The muscles of the face are most frequently affected. A sudden, quick blinking of the eyes, which may be repeated very frequently at short intervals, or occur only a few times during the day, is the form most frequently seen. The eyebrows in other cases are suddenly elevated; the face may be drawn to one side, or the facial muscles of both sides may be affected, producing a sudden involuntary

grin. The tongue may be affected, and a sudden movement as if moistening the lower lip may be so frequently repeated as to produce an inflamed condition of the skin of this area. In one child, who had been able voluntarily to produce a movement of the ears, a sudden jerky movement of both ears developed independently of the volition of the patient. Spasm of the muscles of the neck results in a jerking of the head to one side; shrugging of one or both shoulders is not infrequent. There are usually no other symptoms apart from the motor phenomena. Hysterical outbreaks are usually met with in the type subject to this affection.

**Diagnosis.**—In rare cases several groups of muscles may be affected at the same time, but this can be easily differentiated from Sydenham's chorea by the purposive character of the movements.

There is a condition of the muscles of the face, most frequently localized to the orbicularis palpebrarum and due to the toxic influence of tea and coffee on the nervous mechanism, which should be distinctly differentiated from the habit spasm. The manifestations are lightning-like contractions of the individual fibrillae, affecting all of them in rapid succession, producing at the most a slight quivering motion of the lids, but never leading to the distinct blinking of habit chorea. This is as frequently seen in adult life as in children, and yields rapidly when tea and coffee are excluded from the diet. In some of the cases eye-strain is a factor.

*Habit spasms* should also be differentiated from what has been described as impulsive tic (Gilles de la Tourette's disease). Some of the forms of this condition appear to be a more serious and wide-spread affection of the toxic condition above described, affecting the orbicularis palpebrarum, and due to some intoxication. It is not infrequently fatal. It begins, as a rule, in very early life, although it may occur as late as early adult life. The muscular movements may affect all the voluntary muscles, and are lightning-like in character, with marked fibrillary movements. Another group of cases described under this condition presents the same quick action of the muscles, with mental disturbance and the use of foul language. The explosive, quick character of the movements, the mental disturbance, and the coprolalia should differentiate it.

**Treatment.**—In both habit spasm and impulsive tic an underlying cause should be carefully searched for and removed. In the habit spasm about the eyes, errors of refraction and loss of muscle balance should be first corrected. The mucous membrane of the nose and the condition of the turbinate bodies should be examined to determine any cause for irritation. The ears and teeth should in the same way receive attention in all cases where the symptoms are referable to any part of the face. Irritative reflex disturbances in the genito-urinary tract, such as phimosis, etc., should be relieved.

The general nutrition should be brought to a normal standard and a proper discipline infused in a routine way into the child's life. While punitive disciplinary measures sometimes succeed in early imitative cases, they frequently do harm. The child should be sent to bed at a definite time early in the evening and compelled to remain in bed an hour after the usual time of rising, both as a disciplinary measure and to secure an added amount of rest for the weakened nervous system. A period

of rest in the middle of the day is also advisable. A cold sponge bath or needle bath is valuable as a tonic stimulant if the child reacts well. A simple diet without tea or coffee and with little meat, but with plenty of milk, eggs, and vegetables, is indicated. Alterative tonics, such as Fowler's solution, quinine, and strychnine, are sometimes of value; more frequently better results are secured by the use of bromides and other nerve sedatives. The child should be encouraged to inhibit the movements as far as possible.

**Electric Chorea.**—This is a rare disease, first described by Dubini, and is manifested as intensely rapid rhythmical movements in the extremities, rarely in the head and face. The movements may be very violent, and have the appearance of being produced by an electric shock. In the severer form described by Dubini as occurring in Italy, paralytic symptoms supervene, and may be associated with epileptiform convulsions. Pain in the head and neck may be present early, and toward the end of the attack atrophy and wasting of the muscles may occur. Fever may be present. The cases terminate in a few weeks or months from heart failure or coma. Jaccoud considers the affection to be due to a symptomatic chorea due to a low grade of cerebrospinal meningitis. A form of electric chorea, probably due to hysteria, has been described by Bergeron. Henoeh has also described a form of electric chorea differing from both the above, and manifested by spasmodic attacks of lightning-like contractions confined to the muscles about the shoulder blade; it is probably a form of myoclonus.

**Treatment.**—This should be directed to the removal of any underlying intoxication. Free purgation, chloral, and bromides have been used, but to little effect.

### HUNTINGTON'S CHOREA.

**Synonyms.**—Chronic progressive chorea; chronic hereditary chorea; degenerative chorea.

**Etiology.**—**Age.**—The disease is preëminently, if not exclusively, a disease of adult life. It usually begins between thirty and forty years of age. Premonitory symptoms, in the nature of mild choreic movements, or more frequently in the nature of extreme clumsiness, or sometimes in an extra precision of movement, occur as early as the twentieth year. Cases have been reported in childhood, *i. e.*, in the second decennial period, by Hoffmann, Peretti, and others. Stevens reports a case in which the chorea developed in infancy. The accuracy of this case as one of Huntington's chorea has been questioned. Jolly reported a case in which chorea and epilepsy developed unquestionably at nine years of age. Remak reports a case in which epilepsy was present from the twenty-third to the thirty-first year, followed by chorea at forty. Diefendorf reports a case in which epilepsy developed at seventeen and choreic manifestations appeared at fifty-three.

**Distribution.**—The cases occur practically over all the European countries—with the exception of Turkey—the West Indies, and South America. Several cases have been reported in negro families, associated with other nervous maladies, the most important of which is epilepsy.

**Heredity.**—Direct heredity has been found in practically all of the cases studied. The exact percentage, however, of those affected who developed chorea has not been determined. Huntington states that “if one or both of the parents have shown manifestations of the disease, and more especially when these manifestations have been of a serious nature, one or more of the offspring almost invariably suffer from the disease if they live to adult life; and if by any chance these children get through life without it, the thread is broken and the grandchildren or great-grandchildren may rest assured that they are free from the disease. Unstable and whimsical as the disease may be in other respects, in this it is firm; it never skips a generation to manifest itself in another; as soon as it has yielded its claims, it never regains them.” Huntington further speaks of the presence of the nervous temperament in all or nearly all of the families in which the choreic taint exists, and that “nervous excitement to a marked degree almost invariably attends upon every disease these people may suffer from, although they may not in health be over-nervous.” Heilbroner states that there is a tendency in successive generations for the onset of the disease to be delayed.

**Sex.**—Huntington states that it is more common among men than women. He was not aware that season or complexion had any influence in the matter. Wollenberg has collected statistics of 128 cases in twenty-two families; of which 74 were in men and 54 in women.

**Pathology.**—The picture is that of a chronic diffuse cortical encephalitis. Three of the writer's cases in both the gross and microscopic picture resembled very closely that seen in paresis. There was a marked atrophy of both the convolutions of the frontal lobes and the motor area. The arachnoid was thickened relatively (due to cortical atrophy) and actually, due to a chronic proliferative process. There was a proliferation of the neuroglial network, with an increase of the neuroglial cells both in the cortical and subcortical tissue. The atrophic process was shown by the absence of the tangential fibres and by the marked perivascular and pericellular space dilatations with both perivascular and pericellular neuroglial nuclear proliferation. The ganglion cells of the cortex appeared to be fewer in number, and with atrophic and pigmentary changes. There was an accumulation of subarachnoidal fluid (hydrocephalus externus), which was in all probability secondary to the atrophic processes in the brain. Throughout the basal ganglia extensive areas of perivascular proliferative gliosis were present. In other cases simple hemorrhagic pachymeningitis was found. The pathological changes in the basal ganglia, especially the lenticular nucleus, represent the basis for the choreic movements, the cortical cellular changes that for the dementia. Lannois and Paviot explain the development of the disease upon the hereditary malformation of the neuroglia of the cortex.

**Symptoms.**—The heredity, the chorea, the tendency to insanity, and its development in adult life are the important features. Of the chorea, it may be stated that the movements resemble very closely, yet are distinctly different from those seen in Sydenham's chorea. They are essentially of the same purposeless, jerky nature. There is a gradual development, at first an apparently extra precision of voluntary motion,

followed by a distinct clumsy, irregular action of the muscles. They are in the beginning of minor intensity and may be confined to the upper extremities or the face. Later, the legs, and afterward the voluntary muscles of the entire body, with the exception of the eye muscles, become affected. In its final development there is a tendency to more symmetry of movement than is seen in Sydenham's chorea. This results, when the lower extremities become affected, in a shuffling, dancing type of gait, associated with irregular movements of the trunk and arms. Choreic movements of the facial muscles cause the production of grimaces somewhat allied to those seen in some cerebral types of chorea. Speech, sooner or later, becomes affected on account of the involvement of the tongue, the lips, and the respiratory muscles, and assumes a hesitating, stuttering, explosive type of articulation. The movements, as a rule, but not necessarily, disappear during sleep. The movements are under the voluntary control of the patient, even when moderately severe. A patient, for years under personal observation, although suffering severely from choreic movements, was nevertheless able to attend to his occupation as a carpenter, and to drive nails with fair precision. The motor power is, as a rule, not involved. In terminal cases, a general or hemiplegic loss of power is sometimes noted. The reflexes are exaggerated, as a rule. The electrical excitability remains normal.

**Psychic Symptoms.**—Huntington stated that "the tendency to insanity, and sometimes that form of insanity which leads to suicide, is marked. I know of several instances of suicide of people suffering from this form of chorea, or who belonged to families in which the disease existed. As the disease progresses, the mind becomes more or less impaired, in many amounting to insanity, while in others mind and body both gradually fail until death relieves them of their sufferings. At present I know of two married men, whose wives are living, and who are constantly making love to some young lady, not seeming to be aware that there is any impropriety in it. They are suffering from chorea to such an extent that they can hardly walk, and would be thought by a stranger to be intoxicated. They are men of about fifty years of age, but never let an opportunity to flirt with a girl go past unimproved. The effect is ridiculous in the extreme." Other observers have also noted the mental condition to be a severe and gradually progressive deterioration, ultimately ending in absolute dementia. In some cases the mental defect is noted from the onset of the symptoms; in others, the mentality may remain unimpaired for years. Mental deterioration is the rule, and it is associated with a loss of memory and a tendency to self-destruction which gradually develops. When the mental degeneration is well marked, outbreaks of violence are sometimes noted. In one of the writer's patients, as the disease progressed, the clinical picture of paresis was presented. The chronic delusional state is more often noted than would be inferred from Huntington's description.

**Diagnosis.**—The other forms of adult chorea apart from the infectious toxic variety of Sydenham may occur, but this is the only type which presents any difficulty in diagnosis. The senile chorea associated with arteriosclerosis, and the degenerative processes of the cortex which go

with it, are sometimes mistaken for Huntington's chorea. This condition, however, only appears long after middle life, and lacks the most essential feature in Huntington's chorea, the heredity. In investigating the family history it should be borne in mind that the disease is usually looked upon as a family affliction, and its presence in other members of the family is at first denied by the patient. It requires tact and careful questioning to elicit this important factor in the diagnosis.

**Prognosis and Treatment.**—The prognosis is absolutely hopeless. Nothing has been found to retard or inhibit the course of the disease.

### CONVULSIONS OF INFANCY AND CHILDHOOD.

Convulsions occurring in infancy and childhood vary greatly in nature, in causation, in the clinical picture, and in the resultant effect upon the nervous system. They may present the symptomatic representation of some general disease or some toxic condition or, on the other hand, they may present, as an epilepsy, the disease itself. This article will consider the subject of infantile convulsions as distinct from epilepsy.

**Etiology.—Age.**—In most cases fits occur before the completion of the first dentition. Of 300 cases studied by Cautley,<sup>1</sup> 44 began in the first six months of life, 52 in the second, 76 in the second year, and 44 in the third year. Thus, no less than 216 were infants under three years of age, 27 were in their fourth year, and the remainder under twelve.

There is no question of the different reaction of the nervous system in infancy, childhood, and adult life. Even when there is a definite pathological factor, as disease of the thymus, this variation is shown, for at different periods of the development of the nervous system, thymic asthma, convulsions, and myasthenia gravis are presented. At the onset of the acute infectious fevers the rigor of the adult is sometimes represented by a distinct convulsive seizure. This variation is more one of degree than of essential difference. Too much stress is often laid upon the term "convulsion." While loss of consciousness is the rule in convulsive seizures, it need not necessarily be absolute. The motor intermittent muscular reaction, or a decided chill, which has been described by a sensation of cold, is essentially a convulsive seizure of nervous origin, and not under the control of the patient. The same grade of intoxication in infancy and early childhood produces a more acute motor reaction with disturbance of consciousness.

The work of Soltmann explains to a certain extent this variation. He found in experimental work on animals that the irritability of the motor nerves in the newborn was negative, while the irritability of the sensory nerves was low. Both varieties of nerves after birth, however, not only reach the normal irritability, but actually develop an electrical hyper-irritability above that seen in the adult. The cerebral motor area is undeveloped and does not react at birth to electrical irritation. The reflex inhibitory centres and paths were also found entirely wanting. An important factor is the lack of synchronous development of the

<sup>1</sup> *The Clinical Journal*, 1905.



inhibitory with the motor function and the peripheral irritability. As a result of this the reflex irritability in the newborn is somewhat diminished, and thereafter shows an increase to a point above normal. Soltmann also determined an increased irritability from the fifth to the eleventh month. While later investigations have shown that in certain respects it has been necessary to modify these observations in all cases, they remain for the most part confirmed. All the observers are agreed upon the absence of the inhibitory function at birth, its delayed development, and the incomplete function of the psychomotor centres.

Apart from these factors, the absence in infancy of volitional motor control, that lack of what might be termed subconscious "willing," a part of the psychic function, appears to be an important element in the production of convulsive states. The well-trained and well-disciplined nervous system is less likely to show motor unbalance under stress and insult than when the opposite is the case. In infancy, lack of such training and discipline, together with the failure of synchronous action of the different functional areas of the brain, combined with the varying progress in the development of the elemental functions, renders the nervous system susceptible to disturbances of motor and psychic correlation by toxic, traumatic, or organic influences. In other words, there is wanting not only the automatic inhibition of the nervous system, but also its reinforcement by volitional inhibition and control. The production of convulsive seizures can best be understood in the simple reflex group. Convulsions may be produced in guinea-pigs by irritation of the sciatic nerve. In a patient under personal observation convulsions were produced by the dressing of an irritated surface on the leg. The patient, a man, aged forty-two years, had convulsions during childhood up to the sixth year, and was entirely free until the necessity arose for active treatment of the local condition. On every occasion on which the ulcerated surface was dressed without the use of cocaine, a general convulsion resulted, which could be entirely prevented by proper local anesthesia, and ceased to occur after improvement in the part. In toxic cases the irritation is probably direct upon the central nervous mechanism. In convulsions of gastro-intestinal origin there is in all probability a combination of factors—a reflex exciting factor, a complete toxic factor, and a possible disturbance of the cardiorespiratory mechanism.

**Predisposition.**—The individuality in children, as regards motor balance, coördination of the psychic, motor, and inhibitory functions, and the defence of the body in resisting invading toxins is as varied as the physical character. This, in some cases, is a matter of heredity; in others, and this is more particularly true in infancy and early childhood, hereditary influences do not play an important part. In a family of six children of good heredity one of the children had convulsions at the onset of measles, scarlet fever, and influenza, whereas the other children suffering from the same affections of approximately the same intensity were free from convulsive seizures. We may, therefore, assume even in infancy a predisposition to the convulsive habit. This is true both of the convulsions of a reflex nature as well as those to which the term "idiopathic" has been given. Moon, who made a careful study of

200 cases, found that convulsions in infancy associated with reflex causes were quite as serious as regards frequency and the mental and moral deterioration as those of an essential nature. In all the varieties of the convulsions of childhood a certain intrinsic condition of the brain in the nature of an instability of function is the most important etiological factor.

Convulsions in childhood almost always have a serious aspect. With proper care and attention at the time of the convulsions, and with careful attention to the life, education, and after-treatment, they may mean nothing more than an accidental complication of some underlying condition. On the other hand, they may be the starting point of a serious disturbance in the function of the growing nervous system, with mental and moral deterioration, and neuroses and epilepsy as the end-result.

**Reflex Causes.**—Reflex disturbances, in the writer's opinion, have been much exaggerated as a causative factor. That they are an important determining factor the writer is not prepared to deny. Except, however, for those which produce sufficient sensory irritation to amount to pain, they act, for the most part, in lowering the general health and nutrition, and causing a loss of nerve tone sufficient to bring the nervous system to a convulsive reaction. We should divide reflex factors into those which are purely reflex in their action and those associated with the disturbance of metabolism. One of the most important factors, namely rickets, which has been placed by some authors in the reflex group, probably acts in both ways. Irritation and subcutaneous lesions of the skin, such as that resulting from excessively hot baths, scalding and burning of the skin, sudden and violent variations in the surrounding temperature, painful wounds, and continued irritation from foreign bodies, often act as the determining factor. Other painful conditions, such as herpes vaginalis, phimosis, pressure on the testicles, erosion of the genitals from irritation of urine, etc., may act in a like manner. Diseased conditions from the presence of foreign bodies in the nasal or aural cavities are sometimes responsible. Carious conditions of the mastoid and disease of the middle ear may be associated with convulsions without evidence of sinus or meningeal trouble. Reflex conditions in the gastrointestinal tract have been given as a cause in a large class of cases. The eruption of the teeth is given as an important factor by many authorities. Henke, however, as early as 1818, called attention to the fact that teething was a normal process, and that it usually occurred at about the same time as a rather rapid development both in the brain and in the general economy, and that the resultant increased sensibility was more to blame than the local process. Most observers have supported this position, which also received confirmation from the work of Soltmann. These statements refer to simple teething processes, and not to those cases in which local inflammatory and other complications exist. Ulcerative conditions of the aural mucous membrane, enlarged tonsils, and nasopharyngeal adenoids may act in a purely reflex manner or assist in lowering the general nutrition and nerve tone.

Pathological factors in the stomach and intestinal tract present complicating factors besides the simple reflex action. Overloading the

stomach of a person with a finely balanced nervous system with indigestible food may be the determining factor in a convulsive seizure not only in childhood, but at any period of life. The ingestion of irritating substances, such as alcohol, reacts in a like manner. Vogel describes convulsive conditions as the result of a gastritis from the ingestion of sulphuric acid and caustic potash; Bouchat, as the result of the various catarrhs of the gastro-intestinal tract; Romberg, from the presence of foreign bodies. Intestinal parasites of various kinds have been blamed for the production of convulsive conditions. The number of children with convulsions is relatively small in proportion to the large number who suffer from intestinal parasites. Reflex conditions in the genito-urinary tract have been mentioned. Catarrhal conditions, congenital anomalies, bladder and kidney stones are factors.

**Infectious Diseases.**—Of the infectious fevers, pneumonia, scarlet fever, whooping cough, and smallpox are most frequently associated with convulsions. In the acute exanthemata the convulsion is most frequently seen as an initial symptom, whereas in pleurisy and more particularly whooping cough, it occurs in the course of, or more toward the end of, the disease. Even in the exanthemata, however, it may occur with the eruption, or more often as the terminal phenomenon. The initial convulsion may be quite independent of the intensity of the infection or the degree of fever. In some epidemics convulsions are much more frequent than in others. The initial convulsion occurring during the course of whooping cough is of much more serious import than the initial convulsions of other infections. In addition to the toxic factor, the mechanical obstruction to the return of blood from the brain often leads to capillary oozing or even gross hemorrhage. In influenza a non-suppurating encephalitis has been found.

As to other diseases, convulsions are sometimes present in typhoid fever, malaria, diphtheria, bronchitis, dysentery, cholera infantum, pyemia, and septicemia. In miliary tuberculosis with a meningeal complication, epidemic cerebrospinal meningitis, and typhoid meningitis the convulsions of a toxic nature should be differentiated from those of organic cerebral lesions.

**Drugs.**—Alcohol may act directly when given in overdosage or by ingestion from the milk of the mother. It is unnecessary here to call attention to the overdosage of atropine, santolin, strychnine, etc.

**Emotional.**—Fright or other strong emotional disturbance may be the determining factor. In infancy and early childhood convulsions are sometimes precipitated by a strong emotional disturbance. That we are here dealing with a disturbance of metabolism as well as a sudden shock to the nervous system is evidenced by the production of convulsions in suckling children after strong emotional disturbance in the mother.

**Constitutional Conditions.**—*Rickets.*—That this is an important factor in the production of convulsions in middle childhood cannot be denied. Too much importance has, however, been attributed to it in the convulsions of infancy. It does not, as a rule, appear until the latter half of the first year of life, whereas the most serious form of infantile convul-

sions appears much earlier. The effect of rickets upon the general development and more particularly in retarding the development of the nervous system renders the latter susceptible to peripheral irritation and other complicating factors, owing to lowered nutrition. It is much more common in the form of laryngeal spasm than as a general convulsion.

*Congenital Heart Disease.*—Convulsions not infrequently complicate marked cardiac weakness. These may be due to an anemic condition of the brain, or, more frequently, to long-continued, passive congestion in the cerebral veins. Toxic factors, the result of general disturbed metabolism, may play an important part as in kidney and liver disease.

*Thymus.*—Local or laryngeal spasm and generalized convulsions with a fatal termination have been found at autopsy to be associated with enlargement of the thymus. Some of the cases were associated with rickets. Two theories have been adduced to explain the condition; the toxic theory—the most probable—and the mechanical theory, which explains the condition as due to pressure of the enlarged thymus upon the trachea, lungs, the large vessels, or the pneumogastric nerve.

**Symptoms.**—The clinical picture varies greatly, dependent upon the intensity of the convulsion and the time of life. In all cases we may assume a complete loss of consciousness, although this is difficult to prove in early infancy. The earlier the age at which the convulsion occurs the more of the toxic and the less of the clonic element is present. The relative or complete absence of tonic movements has given rise to the term “inward spasm,” so much in vogue by mothers, although this term is sometimes used to designate the minor laryngeal spasms with slight rigidity. The convulsions vary in intensity from a temporary loss of consciousness with a fleeting spasm, which may be general or local, to a prolonged convulsion with both tonic and clonic movements. In the more severe forms the onset may be sudden, without previous warning, or more rarely there may be prodromal symptoms, which consist of extreme restlessness, irritability, and minor motor phenomena, such as twitching of the hands, feet, eyelids, and face. In the mild attacks a marked pallor of the face is first noticed, the eyes become fixed, at times roll up in the orbit, the breathing is shallow, the body becomes rigid, a few twitchings of the face and the extremities occur, and the attack is terminated by a sudden deep inspiration.

In the more severe forms the *motor* phenomena become more marked. The head is retracted, the body arched into a position of opisthotonos, the hands are clenched, the thumbs are buried in the palms, the elbows are flexed, the legs are rigidly extended, and quick spasmodic movements of the extremities occur. The respiration is shallow, but may be interrupted and spasmodic. The pulse is weak, hardly palpable, varies in frequency, and may be irregular. The contraction of the facial muscles causes a succession of grimaces, and may lead to violent contortions of the features with contractions of the eye muscles. There is some frothing at the mouth, the skin is clammy, but later the face reddens and the collection of mucus in the throat produces a coarse, rattling sound. The bladder and the rectum may be emptied; the urine during and

succeeding the attack is scanty and may contain albumin. Consciousness is lost during the attack, which usually passes over into a stuporous condition, although this latter is sometimes absent. A condition of general relaxation with marked prostration may follow the attack or may be absent. In most cases the convulsions become general, even when they start as local manifestations. It is not infrequent, however, to see cases in which the convulsive movements are localized to one extremity, the face, or the side of the body. These local convulsions apparently have as deleterious an effect upon the nervous system as the generalized forms. Transient paralyses are sometimes seen, and have been ascribed to exhaustion of the cortical cells. Permanent paralysis in one or more extremities is sometimes observed, and is due either to actual hemorrhage into the cortex, or in some cases to areas of encephalitis. Isolated convulsions, as a rule, last for a very short time; clonic movements may, however, persist, and in some cases may be prolonged for half an hour or longer. In exceptionally rare cases, more particularly in later childhood, twitchings of certain muscles may persist as a chronic affection. Successive convulsions may follow each other in rapid succession, and eighty convulsions have been noted in a single day (Holt). One matter ought to be kept in mind with reference to partial convulsions, namely, that isolated or Jacksonian convulsions are not to be considered as evidence of a focal lesion in infancy and childhood.

*Death* may result from a single attack; this, however, is a rare occurrence, and only occurs in very young infants or in those in whom the vitality is very much lowered from rickets or other diseases. Death may, ensue from exhaustion after long or repeated convulsions.

The paralytic phenomenon above mentioned is the most common sequel of a convulsive attack. Postclamptic blindness has been reported in a small number of cases; Cautley found 8 under eighteen months of age, and 3 between two and a half and three years of age. Temporary aphasia was present in one of these. Ten of the eleven collected cases recovered their sight. In none of the cases examined was there any evidence of disease of the optic nerve.

**Diagnosis.**—The diagnosis of a convulsion in infancy and childhood is not a matter for serious consideration. The differentiation into types is also one of relatively little importance. The problem before the physician is as to the cause of the condition and the possibility of its removal. In the consideration of this subject we have purposely omitted convulsions due to gross cerebral disease, but these ought to be kept in mind, and in the event of recurrent isolated convulsions careful examination of the eyegrounds, of muscle balance, of reflexes, and muscular power ought to be made. Convulsions complicating brain diseases without an acute meningitis are not, as a rule, accompanied by a marked rise in temperature. Convulsions immediately after birth, and more particularly after a prolonged and difficult labor, suggest, as a rule, the presence of cerebral hemorrhage. Convulsions usually indicate the onset of some acute disease when they occur in a child over two years of age in association with a high temperature and clinical evidence of some acute infection. A reflex and toxic factor is sometimes here intermingled on

account of the associated conditions of the gastro-intestinal tract, and may to a certain extent cloud the diagnosis.

The diagnosis of reflex convulsions from worms, phimosis, etc., becomes a matter of simple observation. Rickets is frequently overlooked. These conditions should always be kept in mind. In an investigation of what was thought to be epidemic cerebrospinal meningitis in a home for children, those affected were found to be suffering from rickets.

One of the most difficult problems is the differentiation between *eclampsia* and epilepsy. This is a most difficult matter. While it is impossible to diagnose the fits of infancy as definitely epileptic, it is equally impossible to state that repeated eclamptic attacks in childhood will not persist in later life as epileptic attacks. The physician is permitted to state that in a single convulsion or group of convulsions in infants associated with a definite toxic or reflex factor, the presumption is in favor of normal nervous health in the future. If the convulsions are continued in later childhood, even with a reflex toxic or febrile factor, while the probability under proper conditions of schooling, hygiene, etc., may be in favor of freedom from convulsive seizures, serious stress or insult to the nervous system may develop and thus lead to the convulsive habit. The so-called idiopathic convulsions are more likely to be of a true epileptic nature. Convulsions during the first year are more likely to be of an eclamptic than of an epileptic nature. As childhood advances, the probability is more likely to be in favor of epilepsy; this is more particularly true when heredity is bad and there is evidence of a degenerate nervous system, a distinct aura preceding the attack, with an initial cry and a recovery to a relatively normal condition of health immediately after the attack. It is hardly necessary to call attention to the need of a careful examination of the urine to exclude renal disease, or of the blood to exclude anemia or syphilis.

**Prognosis.**—As far as the individual attack is concerned, this must depend upon a study of the pulse and respiration, the length of the attack, the grade of exhaustion, and the degree of cyanosis and stupor. Cyanosis is an important symptom in the convulsions complicating disease of the respiratory tract. In nephritis the prognosis will depend on the severity of the renal disease.

**Treatment.**—This depends upon a study of the causative factors, their removal, and a study of the isolated attack.

Convulsions in infancy and childhood must always be considered as a dangerous emergency condition. There should be no delay in attendance, and no procrastination in active treatment. If the convulsion is of the simple variety, and not dependent upon passive cerebral congestion, the child should be immediately placed in a quiet, darkened room, and the body placed in a mustard bath if easily accessible, or, what is more usually convenient, a mustard pack. The combination of a mustard pack and a hot foot bath, continued until there is distinct red skin reflex, will often be found to be all that is necessary in the mild convulsion. If the convulsion does not rapidly subside, the inhalation of chloroform should be immediately begun. If there is a history of constipation or overfeeding, the lower bowel should be emptied by a simple enema,

or in older children by sodium or magnesium sulphate. Immediately following the emptying of the bowel, chloral and bromide should be injected. Three or four grains (gm. 0.25) of chloral and from 5 to 10 grains (gm. 0.3 to 0.6) of potassium bromide should be given to a child aged six months, and repeated in an hour if necessary; in older children a larger quantity in proportion may be administered. The injection should not exceed one to two fluidounces in bulk (30 to 60 cc.), should be injected high, and prevented from escaping by tight pressure on the buttocks. This can be repeated within an hour if necessary. If the injection is not retained, or if the convulsion should continue in spite of it when the chloroform is withdrawn, morphine should be given hypodermically (gr.  $\frac{1}{40}$  at six months, repeated in an hour if necessary, gr.  $\frac{1}{20}$  at one year, gr.  $\frac{1}{16}$  at two years, with an increase in this dosage if the occasion requires it). If the attack has subsided, the child should be kept in a quiet room for a few days, the bowels should be thoroughly evacuated, a light diet should be administered, and small dosage of bromides given for at least a week. If the convulsions show any tendency to recur, antipyrine may be combined with the bromides. A hot bath or a hot pack is contra-indicated in convulsions complicating advanced pulmonary disease, with extensive disease or collapse, or when the fit is due to syncope. Careful search should be made of all the possible sources of reflex irritation, and these should be immediately corrected. The child should be carefully watched, and its health kept up to a normal standard.

## CHAPTER XX.

### MYASTHENIA GRAVIS. PARAMYOCLONUS MULTIPLEX. PERIODIC PARALYSIS.

By DANIEL J. McCARTHY, M.D.

#### MYASTHENIA GRAVIS.

**Synonyms.**—Myasthenia gravis; pseudoparalytica; ascending bulbar paralysis; asthenic paralysis; pseudobulbar paralysis without an anatomical pathological basis.

**Definition.**—A disease with fatigue symptoms referable to the muscular system, due to an exhausted condition of nervous enervation, without definite pathology in the nervous system, and with minor changes (lymphocytic infiltration) in the muscles.

**Etiology.**—In the preparation of this article 180 cases were extracted, and two unrecorded cases added from the author's case histories.

**Sex.**—There were 83 males and 96 females. The preponderance of the latter can be accounted for by the influence of pregnancy, the menopause, and the puerperal state.

**Age.**—The disease is most frequent in the third decade.

Age.	Males.	Females.
Under 10 . . . . .	2	2
10 to 20 . . . . .	8	18
20 to 30 . . . . .	21	42
30 to 40 . . . . .	18	20
40 to 50 . . . . .	24	5
Over 50 . . . . .	9	3

The average at which women are affected is decidedly below that for men. As will be seen, males are more frequently affected during the fifth decade, women during the third.

**Occupation.**—The station in life and occupation have no decided influence on the disease.

**Infectious Diseases.**—Acute inflammatory processes occur with sufficient frequency immediately before the disease as to constitute, if not an important etiological, at least an important determining factor. If we consider all the possible sources of infection, including the nasopharyngeal infections, we find in our tables 39 cases. Influenza was present ten times, and among the other infections, scarlet fever, typhoid fever, diphtheria, syphilis, herpes, puerperal and postpuerperal infections were noted.

**Nervous Diseases.**—The consideration of this disease by neurologists as a nervous disease has led to a very careful study of possible nervous influences. A distinctly neurotic history, however, is present in only a relatively small number of cases. The history of nervous disease in some



member of the family was present in 35 cases. In most of these the types of nervous disease in parents or other members of the family were of such a nature that they could easily be excluded as having a distinct bearing upon the development of the disease. In 12 of these cases, for example, it was stated that there was a family history of "nervousness" without having any distinct statement as to the meaning of this term. The mother of one patient suffered with unilateral ptosis, the mother of another showed a defect of the ocular muscles, and migra ne was present in still another case. Oppenheim has paid particular attention to evidence of a defectively constituted nervous system. Syphilis and alcoholism may be considered as negligible factors, the former occurring in 2 cases, the latter in 1. Pregnancy and puerperal conditions were present in 11 cases. In 2 of the writer's cases a miscarriage was regarded as the causative factor. In most of the recorded cases pregnancy has caused a distinctly deleterious influence on the course of the disease. In one recorded case, however, the patient felt very much improved during the pregnant state, but she was much worse during lactation. The menstrual period has a similar effect; patients are decidedly worse at this time.

**Pathology.**—The changes are mainly confined to the muscles. The nervous system in the vast majority of cases is entirely normal. Weigert, in a case of myasthenia gravis, which presented lymphosarcoma of the thymus gland, found at autopsy an accumulation of small round cells in the endomysium and perimysium, and considered them as metastasis from the thymus tumor. In subsequent reports these accumulations of cells have been termed lymphorrhages, and inasmuch as they may occur without primary malignant disease, are looked upon as lymphoid structures. They may be present when the thymus rest is normal. Examination of a muscle of an affected case will show the muscle fibres to be perfectly normal. Within the area of lymphoid infiltration, however, a few fibres may be atrophied and lack striation. A lymphorrhage or lymphoid infiltration occurs as microscopic accumulations of an irregular shape between the fibres, and only rarely infiltrating them. They consist for the most part of small lymphoid cells from one to one and a half times the diameter of a red blood cell. This smaller type of cell resembles the cells of the thymus, but is usually larger than the small cells of the fetal thymus. A few cells of a larger type, containing a relatively larger amount of protoplasm and a small nucleus, are occasionally seen. Polymorphonuclear leukocytes are only exceptionally present. A loose reticulum is present between the cells. The composite structure closely resembles adenoid tissue, and the term "lymphorrhage" is an appropriate one. Capillary hemorrhages are not infrequent in the neighborhood of the lymphoid accumulation.

When these changes occur they are constant in affected muscles. Of the 29 autopsies reported since they were first described, there is a record of examination of the muscles in 16 cases, and lymphorrhages were present in 12 of these. Abnormalities of the thymus cannot be considered as a constant feature in this disease. Lymphoid deposits occur when the thymus is normal; they cannot, therefore, be considered, at least in all cases, as secondary to thymus disease. The presence of disease of the

thymus gland in 10 cases of the 180 reported, while not a constant factor, yet occurs sufficiently frequent to be considered in a causal relation to at least one group of these cases. Of the 10 cases recorded, simple persistent enlargement of the thymus was found in 4 cases (in 2 of these associated with hemorrhages and an absence of eosinophiles in the thymus). Lymphoid enlargement of the thymus was found in 4 cases and abscess of the thymus in 2 cases. Malignant disease elsewhere in the body was found in 4 cases.

**Nervous System.**—The examination of the central and peripheral nervous system has given, as a rule, negative results. The presence in a small number of cases of slight degenerative changes in the ganglion cells and capillary hemorrhages in the cerebral gray matter have been considered as agonal. Lymphorrhages in the posterior root ganglia in one case may be considered as accidental as regards the location in the nervous system, and in no sense concerned in the production of the disease. From both a pathological and clinical standpoint the presence of lymphorrhages in otherwise normal muscles presents a valuable means of diagnosis in a suspected case. After extensive control work they have been found in other conditions only on one occasion, in a case of amyotrophic lateral sclerosis. In this disease there is, however, an associated degeneration of the muscle fibres, with a degeneration of the lateral tracts and gray matter of the spinal cord. The more recent work supports the statement of Buzzard, that his "experience affords reasonable grounds that lymphorrhages are constantly present in the muscles and other organs in cases of myasthenia gravis." Their detection, however, entails a most thorough and diligent search of large numbers of sections.

Much speculation and relatively little accurate knowledge as to the cause of the disease is to be found in the literature. The consensus of opinion appears to be that the disease is due to some toxic agent. Weichardt has shown that physical fatigue in the normal individual is caused by the production of a toxin in the muscles. This toxin, however, cannot be demonstrated in the circulatory blood. If the toxin which he has isolated be injected into animals, an antitoxin is produced which is found in the circulating blood and acts *in vivo* and *in vitro*. He has also shown that the toxin of excessive fatigue may cause death. It would appear that the preservation of muscle excitability is dependent upon a period of rest either for the removal of the toxin from the muscle, or the production of an antitoxin in the blood which counteracts the fatigue toxin. Inasmuch as this toxin cannot be demonstrated in the circulating blood, the method of its removal from the muscle is not altogether clear. Link believes that the lymphorrhages in the muscles interfere with the lymphatic circulation, and thus prevent the removal of the fatigue products. This theory assumes the presence of lymphorrhages in the muscles early in the disease. Another view is that the toxin is not due to a disturbance of metabolism, but is of an exogenous nature, the result of some bacterial infection. The supporters of this view point to the presence of an infectious process in the history of a large percentage of cases, and to the fact that the infections are such, scarlet fever, typhoid fever, influenza, etc., as are not infrequently followed by a toxic neuritis.

The presence of the *myasthenic reaction* is the best evidence that the toxic action affecting the nervous system is not restricted to the upper motor neuron. The fatigue exhaustion produced by the faradic current and its similarity to that produced by voluntary muscle exercise would point to the muscular or intramuscular neuromechanism as the seat of derangement. It would appear from the experiments of Buzzard that the myasthenia is due to an exhaustion of the nerve end organs. In a well-marked case of myasthenia gravis, a moderate galvanic current was applied to the biceps muscles and a contraction obtained. The muscle was then faradized until it gave no response to a strong stimulus. Then it was tired out by making the patient flex the elbow against resistance exerted until all power of flexion was lost. On applying the same strength of galvanic current as used at first, an excellent contraction was obtained. Finally, on again applying the faradic current the muscle was found to be still unresponsive. The peculiar muscular phenomena in this disease, such as are demonstrated by this test, can perhaps be best explained by the work of Botazzi, confirmed later by Iotzko. They hold that in the muscles there are two contractible substances, a fibrillar and protoplasmic, giving different electrical reactions. The fibrillar substance gives a quick, lightning-like response, such as is produced on the normal muscle by the make or break of the galvanic current, whereas the reaction of the protoplasmic substance is slow and such as is seen in a degenerated muscle. Both substances react to the galvanic current, but the protoplasmic substance requires a much stronger stimulus than the fibrillar. A selective poison acting more on the protoplasmic and reducing its excitability, would explain the exhaustion after a faradic or voluntary fatigue, whereas the muscle would still react to the galvanic current by the stimulation of the fibrillar substance. This theory relieves the peripheral nervous system of much of the responsibility that has been placed upon it in the production of myasthenia gravis.

Assuming the disease to be of toxic origin, a very natural query is as to its nature. The view above stated as to the exogenous origin as the result of bacterial invasion has little to support it other than the presence of an infectious process at the onset in thirty-nine cases, and an analogous condition as the result of a toxin in diphtheria and influenza.

The theory of an endogenous toxin, originally proposed by Weigert, has been supported by investigations in the pathology of the disease. He held that in Graves' disease and Addison's disease a definite internal secretion was necessary to the maintenance of bodily health; persistence of a secretion from a gland (the thymus) whose function in a normal individual should be in abeyance likewise gives rise to metabolic intoxication manifested by myasthenic symptoms. This theory, of course, applies to the group of cases in which the thymus was found diseased. It would not apply to the cases in which the thymus rest is found in a normal condition. A large number of cases in which disease of the thymus is not present may be explained (assuming that the thymus is a lymphatic structure) by possible lesions of the lymphatic system elsewhere than in the thymus gland. This is supported to a certain extent by the presence of lymphoid collections (lymphorrhages) in the heart,

liver, and kidney. It is of interest to note that an identical syndrome to that of myasthenia gravis is seen in cases of myeloma in which the bone marrow is extensively involved (Senator).

As the lymphorrhages are not present in the affected muscles in a sufficient number of cases to account for the disturbance of function, we are forced to regard them as the physical manifestations of some underlying intoxication of unknown origin.

**Symptoms.**—These are almost entirely confined to the muscular system. After a single muscular effort the muscles show decided fatigue, and, if the effort is continued, paresis, or even complete paralysis, results. After a period of rest the muscle returns to its normal condition. After a night's sleep the affected muscles may present a relatively normal condition; but, with the fatigue incidental to a day's routine, a moderate or even a marked degree of weakness is presented. This condition is most marked in muscles such as the elevators of the eyelids and the muscles of the jaws, which are maintained in more or less constant action during waking hours. The voluntary muscles throughout the entire body may be affected. The muscles supplied by the bulb, however, are those which are most frequently and earliest involved. The symptoms may be restricted to this group of muscles throughout the course.

**Bulbar Muscles.**—The disease, as a rule, is of slow onset, and begins with an involvement of the levator palpebræ superioris. The lids show a tendency to droop as the day progresses, until at nightfall the eye may be entirely closed. While ptosis, as a rule, is bilateral, it is not unusual to find it more marked on one side. Until the disease is well advanced, the drooping of the lids is entirely or almost entirely absent after a period of prolonged rest. In the advanced cases a well-developed or even complete ptosis may be present even after sleep. The other eye muscles, both intrinsic and extrinsic, the orbicularis palpebrarum, and the occipitofrontalis may also be affected.

**Diplopia** due to weakness of the external ocular muscles is a frequent symptom. It was noted as an initial symptom in thirty-one cases. The type of diplopia presented is fairly characteristic. In successive examinations, even at short intervals, there is a marked variation in the position of the images. This is due to the varying type of palsy as a result of the exhaustion of the different eye muscles during the progress of the tests. The weakness of the eye muscles and the relative loss of muscular balance give rise to irregular, jerky movements when the eyes are deliberately moved in one or another direction during examination. This condition has been mistaken for a true nystagmus. The convergent movement of the eyes, although at first fairly good, is easily exhausted. As a result of this, reading becomes impossible on account of the blurring of the print. The inequality of the pupils and a loss of reaction of the pupils to light has been noted in a few cases. A sluggish reaction of the pupils to light is of more frequent occurrence. The sphincter iridis does not show the fatigue reaction to repeated light stimuli. Hippus was recorded in one case but no significance can be attached to it. The involvement of the upper muscles of the face, the occipitofrontalis and orbicularis palpebrarum, in association with the ptosis gives rise to a fairly characteristic position of the

head. An isolated bilateral ptosis is partially or completely compensated for by contraction of the occipitofrontalis muscles. When this latter group of muscles is also affected, the advancing ptosis can only be compensated for by throwing back the head in an attempt to see under the lids. When the muscles of the neck become involved, it is impossible to maintain this backward position of the head, and it then not infrequently takes a forward paralytic position, resting on the chest.

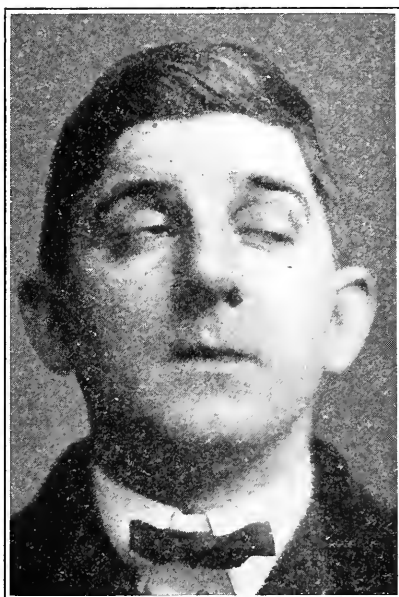
Other ocular symptoms have occasionally been noted. Gunn, in one of Buzzard's cases, noted an associated dilatation of the pupils with elevation of the lids under emotional stress, although the patient was not able to

FIG. 45



Myasthenia gravis. (Case of Spiller and Buckman.) The ptosis of the right eye is not so great as that of the left. The photograph was taken one or two minutes after the glasses were removed. At times he is able to open both eyes fully.

FIG. 46



The eyeballs are almost completely covered. The photograph was taken after the glasses had been removed three or four minutes.

deliberately elevate the lids by a voluntary effort. von Graefe's symptom has been reported in several cases, and Stelwag's in an isolated case. Both these symptoms are not infrequent in tuberculosis and in exacerbations of chronic alcoholism. They may be considered as significant of an underlying intoxication.

Involvement of the muscles of the lower *facial* distribution results in a loss of expression, an inability to pucker up the lips or whistle, a drooping of the angles of the mouth, drooling, and the so-called "nasal smile."

The muscles of mastication and deglutition, including the tongue, may be affected relatively early, or the involvement may follow in order of regular progression after the eye and face muscles. It was present at the

onset of the disease, in association with eye symptoms, in four cases. The power of mastication may be slowly lost, as during the progress of a meal or it may survive only the first attempt at chewing the food. The loss of power of the tongue may interfere with the transmission of the bolus of food to the pharynx. When the muscles of the palate are involved, liquid food may regurgitate through the nose. In advanced cases of myasthenia of this group, weakness becomes continuous throughout the entire day. When the power of deglutition is retained, the power of mastication can be assisted by the use of the hand. Clonic movements of the muscles of the jaws are occasionally noted. The difficulty in deglutition may become so serious as to necessitate the use of the stomach tube or alimentation by rectum. Choking attacks are not infrequent, and may be a cause of death either from interference with the respiration or from exhaustion.

The laryngeal muscles show fatigue manifestations similar to those noted in the extra-ocular muscles. The laryngoscopic picture is not at all constant, and varies from time to time. The pharyngeal reflexes are usually absent. Anesthesia of the pharynx and larynx has been noted in two cases. Fatigue manifestations in the tongue are associated with fibrillary tremors. The speech is usually indistinct, and when fatigued has the nasal, mumbled character of true bulbar paralysis. In moderately advanced cases there is a paretic element which is not seen in true bulbar cases. The speech fatigue is accentuated in cases with involvement of the muscles of respiration.

Involvement of the chest muscles becomes a very serious symptom. The respiratory excursion is very markedly diminished. Sudden attacks of dyspnoea often terminate the life of the patient. They occur usually toward the end of the day, or after exertion. It is not, however, always a fatigue dyspnoea, and may occur suddenly without any apparent cause. Strümpell considered a paralytic condition of the tongue to be an important factor in one of his own cases, and relieved the condition by pulling the tongue forward. Pneumonic infections are not infrequently a cause of death. Cardiac distress may be present in association with or independent of dyspnoea.

The other muscles of the trunk may become so weakened as the disease progresses as to render the patient unable to sit up in bed, or the terminal paralysis may render the patient completely helpless.

**Extremities.**—These may become involved early, may remain free throughout the course of the disease, or be involved in the general paralytic phenomenon. The muscles nearest the trunk are the earliest affected, although the reverse may be true. When the hand muscles are involved, the handwriting shows the characteristic fatigue and a rapid loss of regularity in letters as the writing progresses. In the lower extremities the fatigue manifestations are usually noticed first in the iliopsoas and quadriceps muscles. Giving way of the legs may be a relatively early symptom. While the patient at first is able to walk fairly long distances, this power is rapidly impaired, until finally there is inability to walk at all. In relatively early cases the fatigue manifestations disappear after a short period of rest.

**Myasthenic Reaction.**—The electrical examination of the muscles gives important data. If a rapidly intermittent current be applied to the affected muscles, there is at first a good, strong contraction; this contraction, however, is not maintained as in a normal muscle, but rapidly decreases until there is no response. If the current is intermitted for a short period of time (from one to two minutes), a strong response is again obtained, which disappears, however, more rapidly than the first. This test is called the *myasthenic reaction*. The rapidity with which the muscle becomes exhausted depends to a degree on the myasthenia present in the affected muscle. In advanced cases, the exhaustion may be complete within twenty to thirty seconds. In early or mild cases, only a relative degree of exhaustion may be obtained, even with strong currents. The reaction is not necessarily constant, and may be different from time to time. It is influenced by the degree of fatigue in the muscles examined. It may be present in muscles supplied by one branch of a nerve and absent in other muscles supplied by the same nerve (Goldflam). After exhaustion of the muscles by the faradic current, a fairly good contraction may still be maintained by voluntary effort. The return of irritability after exhaustion is much more rapid than in normal muscle exhausted by fatigue (Murri).

The reaction to the galvanic current shows only a relatively slight degree of diminution of contraction. The exhaustion of the muscle is not here produced as with the use of the faradic current. Atrophy of the tongue was found in five cases. In ten cases a relatively slight degree of atrophy was noted in the affected muscles.

**Reflexes.**—The reflexes of the extremities, as a rule, are exaggerated. Exhaustion of the quadriceps muscle either by fatigue or by faradization has no effect on the knee-jerk. Ankle clonus is never present, and the plantar reflex was uniformly of a flexor nature. In advanced cases, the jaw-jerk is usually absent. The skin reflexes are normal.

**Negative Symptoms.**—There is no disturbance of sensation. After exercise a sensation of stiffness or aching in the limbs may be present. This sensation is also present in advanced cases due to inability to move the affected parts. In rare instances the muscles may be sensitive to deep pressure. Buzzard reports a case of anesthesia of the trunk, which he ascribes to lymphorrhages in the posterior spinal ganglia.

There is, as a rule, no atrophy or fibrillary tremors of the affected muscles (exceptions noted above). There are no trophic changes in the bones or skin. The visceral and rectal sphincters are never involved.

Mental depressive states have been noted in two cases.

**Urine.**—Spriggs found that in a case of myasthenia gravis the creatinin output, both absolutely and relatively to the total nitrogen, was definitely diminished, while that of uric acid remained normal.

**Diagnosis.**—This should never be a difficult matter if the regular symptoms are kept in mind. A combination of motor symptoms, progressive weakness and rapid fatigue of the voluntary muscles after exertion, and particularly the muscles supplied by the bulb, the presence of the myasthenic reaction and the absence of wasting, fibrillary tremor

or sensory disturbance, with a retention of bladder and rectal control; of slow, more or less interrupted onset, running a variable course with a tendency to improvement and relapse, constitutes a characteristic symptom complex which cannot well be mistaken. The conditions from which it must be distinguished are true bulbar palsy, pseudobulbar palsy, diphtheritic paralysis, polienccephalitis superior, the muscular dystrophies, hysteria, and general asthenia.

*True bulbar paralysis* affects the lower distribution of the face, the tongue and the muscles of deglutition, by preference; the upper facial distribution and the ocular muscle are rarely affected, and then only in the advanced stages of the disease. There is marked atrophy, fibrillary tremors, and the absence of the myasthenic reaction. In *pseudobulbar paralysis*, the upper facial distribution is not affected, and the history of repeated apoplectic attacks will make the diagnosis clear.

*Diphtheritic paralysis* is of more rapid onset, and there is a history of the throat infection. The electrical reactions in the extremities will easily differentiate this from myasthenia gravis.

In *polienccephalitis superior* the onset is sudden. The headache, fulness in the head, and other symptoms of an inflammatory type are associated with constant paralytic phenomena of the ocular muscles, which bear no relation to fatigue.

The facio-scapulo-humeral type of *muscular dystrophy* may bear a striking superficial resemblance, but the presence of muscular dystrophy in other members of the same family, the rapid wasting, the relative freedom of the muscles of deglutition, and absence of the myasthenic reaction should make the diagnosis easy.

In *hysteria* associated with neurasthenia the fatigue symptoms may be so marked as to bear a striking resemblance to myasthenia gravis. Sooner or later transient sensory disturbances, reversal of the color fields, headaches, and other painful phenomena, all of which may be influenced by suggestion, will lead to the diagnosis.

**Prognosis.**—This is unfavorable in the majority of cases. The disease must always be considered as a serious affection, with a possible fatal termination. Of the 180 collected cases 72 proved fatal. These statistics are, to a certain extent, misleading on account of the tendency in case reports to consider only fatal cases. Of the 5 cases which have come under the writer's observation, only the 2 which resulted fatally were reported. On the other hand, there is a disposition to report cases after a short observation without waiting a reasonable time to follow the course of the disease. In the fatal cases, the duration varied from one and one-half to two years. The disease may run an acute course, and in one case terminated in fourteen days. In another recorded case the patient lived fifteen years. It is impossible to state the percentage of cases that go on to permanent recovery. It is probable, however, that a permanent cure often takes place. Respiratory failure with attacks of dyspnoea should be considered a grave symptom. Sudden death in such cases is not of infrequent occurrence. The disease varies markedly in its course and in its symptoms. Even in apparently favorable cases unaccountable relapses are likely to occur.



**Treatment.**—All possible infectious or toxic causes should be, as far as possible, removed. In puerperal cases, septic or toxic processes in the genito-urinary apparatus should receive particular attention. The advisability of the removal of tumors must depend upon the particular case and the location of the tumor.

A full or modified form of *rest* treatment is indicated. The whole therapeutic régime should be so directed as to eliminate all possible muscular fatigue. The diet should be so arranged as to secure the best nutrition with the least effort in mastication, and in severe cases a full milk diet is indicated. In the earlier cases food should be so prepared as to avoid the necessity of mastication fatigue. If the use of the stomach tube should at any time be deemed necessary, either for purposes of diagnosis or alimentation, the greatest care should be used. Death may result from the fatigue and excitement incidental to the use of the tube, or as the result of attacks of choking. Unless the tube is taken easily and without excitement or gagging, its use should be avoided.

Care should be taken to properly protect the patient from cold. In those cases in which the symptoms are aggravated or induced by cold weather, a warm, equable climate should be recommended. Electricity should only be used for purposes of diagnosis, and even then only with the greatest care. Faradism as a therapeutic agent does harm, and galvanism does no good. When massage is used, it should be given only for a very short period of time and the effects carefully noted. It is likely to do more harm than good. The glandular extracts (suprarenal, thyroid, and thymus) have been tried without appreciable results. General tonics would appear to be indicated, and in conjunction with hygienic measures have given good results. Arsenic in the form of Fowler's solution, the iron preparations, and strychnine in small doses have been recommended. Strychnine has been used in massive doses without distinct benefit.

### PARAMYOCLONUS MULTIPLEX.

This is an affection first described by Friedreich in 1891, and characterized by clonic, lightning-like contractions occurring either constantly or in paroxysms, affecting at times all the muscles of the body with the exception of the eye muscles, but more frequently the muscles of the lower extremities.

**Etiology.**—The cases reported have been chiefly in males. Some emotional disturbance, such as fright, is often the determining factor. In pure cases hyperirritability of the nervous system is often present. Tuberculosis has been reported as present in certain cases.

**Pathology.**—Of the three cases which have come to autopsy, the case of Hunt showed hypertrophy of the muscle elements. Friedreich stated that the condition was due to an increased irritability of the gray matter of the spinal cord with an irregular discharge of motor impulses. That the disease is one of the lower motor neurons or of the muscles is accepted. Irritation of the motor cortex does not give isolated muscular movements. Localized contractions of muscles or individual fibres of muscles have

always been considered as having a localization in the gray horns of the spinal cord. We would, therefore, conclude, in view of negative findings in the nervous system, that the disease is the result of some disturbance of function of the inferior neuromuscular mechanism.

**Symptoms.**—All of the muscles of the body may be affected. The muscles of the extremities, and especially of the more proximal portion, are those most usually affected, in the upper extremities the biceps, triceps, and supinator longus, and in the lower extremities the internal vastus, external vastus, the rectus, and the adductors. The muscles affected display the greatest activity, the individual muscles in use springing forward in contraction with the greatest rapidity, as if suddenly irritated by galvanic shock. These contractures are not associated, as a rule, with fibrillary tremor. They do not usually produce any motor response in the extremities. Occasionally a barely perceptible involvement of the arm may be the result of the more violent contractions. The intensity and rapidity of the contractions may be increased by mechanical irritation of the skin, mechanical irritation of the muscles, exposure to cold, and by excitement. They are usually decreased by voluntary motion, although the contrary may be true; they usually disappear during sleep. Occasionally, as in Friedreich's case, the patient may be frightened out of sleep by a sudden very painful crampy movement of both legs, resulting in a sudden jerking of these members against the abdomen. The electrical irritation of the muscles is normal and the reaction of the muscles to mechanical irritation is not increased. There is no disturbance of sensation, no atrophy, and no cerebral, spinal, bladder, or rectal symptoms.

**Diagnosis.**—The symptomatic picture of a well-defined case of paramyoclonus multiplex is distinctive. This term has been used so frequently, however, to designate other forms of myospasm, that considerable difficulty has been experienced in limiting the disease as a clinical entity. There is no question that most of the cases reported in the literature should not be considered as such in the strict sense of the term. It should be remembered that a myokymia or contraction of individual groups of muscular fibrillæ is not uncommonly present in neurasthenia and other functional nervous disorders. The fibrillary tremor of wasting muscles need only be mentioned in this connection.

Dana, in 1903, differentiated the cases recorded as paramyoclonus multiplex into the following groups: (1) Paramyoclonus multiplex of Friedreich; (2) myoclonus of the functional or hysterical type; (3) myoclonia of the convulsive tic type; (4) myoclonia of the degenerative chorea or familial, or myoclonia epilepsy type; and (5) myoclonia of the infectious and symptomatic choreas.

There should be no difficulty from a careful study of the case in excluding cases of the second group. The contractures are not strictly fibrillar, but are of the coarse, irregular, clonic type, associated with minor or more extensive movement of the extremities. There are other evidences of hysteria usually associated, and this is more particularly true in the early stage. The muscular phenomena are not constant, and the frequency is influenced by excitement, excessive attention, etc.

The cases belonging to the third group (convulsive tic type) are easily differentiated as representing a disturbance of cortical origin as differentiated from the spinal type seen in paramyoclonus multiplex. All movements belong to the purposive group of gross movements. There are no fine, fibrillar tremors of the muscles, but a quick, jerky contraction of the muscle groups, such as blinking the eyes, screwing up the face, twisting the head, shrugging the shoulders, etc. In Tourette's disease (*maladie de tic convulsif*) this purposive type of muscular spasm may be widespread and affect the muscles over the entire body.

The myoclonus epilepsy of the fourth group is easily differentiated from paramyoclonus multiplex by the presence of epileptic attacks. It is a manifestation of excessive functional disturbance of the cortex. The movements do not resemble paramyoclonus multiplex so much as they do those seen in Sydenham's chorea. They are, however, prone to be rhythmical and associated with some fibrillary tremors. From the infectious chorea group, this disease is easily differentiated by the gross purposeless character of the movements in the latter disease. From the electrical chorea of Dubini it is distinguished by the presence of the severity and fatal character of the latter affection. In Dubini's disease the rapid rhythmic movements are very violent, as if produced by an electric shock, and may be associated with fever, paralytic symptoms, epileptic convulsions, and atrophy and wasting of the muscles.

**Prognosis.**—This must be considered unfavorable in true cases of paramyoclonus multiplex. The cases reported in which such startling curative results were obtained by the use of electricity are usually looked upon as cases of hysteria. The disease usually lasts a long period of time, unless, as not infrequently happens, the life of the patient is terminated by some intercurrent affection such as pulmonary tuberculosis.

**Treatment.**—Careful attention should be paid to associated visceral disease, more particularly of the lungs. If the patient is under weight, a modified rest treatment with overfeeding is indicated. Careful attention should be paid to keeping the gastro-intestinal tract in good condition. The urine and stools should be carefully watched in order to insure proper elimination through these channels. Flushing of the system by the use of large quantities of water in the dietary and by the occasional use of Epsom salts will be found in some cases to be of assistance. The bromides, chloral, and the valerianates have been used for their calming influence. Arsenic is of value in some cases. Thyroid extract has been used in this disease, but its value has not been established. Various forms of electrical treatment have been lauded as curative agents. The galvanic current, the static spark and breeze have been reported as producing valuable results. They at least offer a helpful means of eliminating the excess of functional symptoms present in most of these patients.

### PERIODIC PARALYSIS. FAMILY PERIODIC PARALYSIS.

**Definition.**—A flaccid paralysis affecting the muscles of the trunk and the extremities, temporary in character and recurring at more or less regular intervals.

**Etiology.**—Judging from the list of cases reported in the literature, this is a rare disease. Most of the cases reported have been in family groups; 5 cases have been reported by Cousot, 22 by Goldflam, 2 by Hirsch,<sup>1</sup> and 11 by Taylor. Mitchell<sup>2</sup> reports one group in which 4 and another in which 5 were affected. Holtzapple<sup>3</sup> reports 17 cases.

That *heredity* is an important etiological factor in these family groups, there is no question. In Holtzapple's series, cases in four generations in the same family were studied. While a neurotic basis for the disease is generally denied, migraine was present in 10 of the 17 cases reported. Five had attacks of paralysis and headache. Thirteen other members of the family suffered from headache. In some of the cases the headaches preceded the attack, developed in early childhood, and disappeared with the development of the paralytic phenomena. In other cases the headaches continued, alternating with attacks of paralysis. This, however, is the only group of cases in which migraine is of etiological importance.

The disease may develop as early as the fifth year, but, on the other hand, may not appear until after thirty.

Transmission may occur either through the male or female. Individual members of the family may be entirely free. These may, however, transmit the disease, the affection in this manner skipping a generation. In Holtzapple's cases, migrainous headaches were present in members of the family who did not have the disease. In this very interesting family the disease seems to have started from the father of the first generation, who suffered from periodic sick headaches. The statement of Taylor that these cases have occurred in families of unusual nervous stability and that in no case do we find evidence of a degenerative family is not borne out. The attacks in some of the cases were most frequent during the cold months. This is of especial interest in connection with the article of Rich,<sup>4</sup> who described a condition of tonic spasm of the face muscles in five members of his own family, due to exposure to cold or dampness, in one case there being a complete general paralysis with the exception of the tongue, as the result of sleeping in moist underclothing. While the paralysis, being of the nature of a spasm, is different from the flaccid palsy of periodic paralysis, the influence of cold in its production is of interest. Sinkler<sup>5</sup> also reported a sporadic case of recurrent facial paralysis extending over a period of ten years, without such etiological factor. Muscular exertion, lack of exercise, indiscretions in diet, exposure to a draft, menstruation, constipation, nervous or mental fatigue, worry, and excitement have been given as conditions which precipitate an attack.

**Pathology.**—Two of the patients in Holtzapple's series have come to autopsy but no lesion which could be associated with the disease was found (Winternitz). Small portions of muscles have been excised and studied by Goldflam and Oppenheim. The former found a waxy degeneration to which he attributes no importance. The latter found an increase of the diameter of the individual muscle fibres, hypertrophy of the fibres, rarefaction of the primitive fibrillæ with vacuole formation. To these

<sup>1</sup> *Deut. med. Woch.*, 1894, No. 32, p. 646.

<sup>3</sup> *Jour. Am. Med. Assn.*, 1905, xlv, 1224.

<sup>5</sup> *Jour. Nerv. and Ment. Dis.*, 1898, p. 744.

<sup>2</sup> *Brain*, 1902, xxv, No. 1, p. 109.

<sup>4</sup> *Medical News*, Phila., 1894, xv, 210.

changes Goldflam attributes qualitative, electrical, reactive changes which he alone found in the free intervals between the attacks. These changes have a special significance when studied with the cases of Bernhardt, which showed from early childhood a persistent and constant loss of power in certain muscle groups, separate and apart from the periodic attacks which develop later. Both of these authors would place the disease on the basis of the muscular dystrophies.

In the absence of any such condition in the other cases reported, the prevailing idea is that we are here dealing with an auto-infection of unknown origin, associated with a lowered condition of metabolism and induced by excessive muscular activity, followed by a period of rest. Extensive investigation of the urine and the blood has not given much information as to the nature of this intoxication. Goldflam and Taylor reported a lymphocytosis with a moderate eosinophilia. In Mitchell's case a study of the toxic effect of the blood serum by intravenous, intraspinal, intrameningeal, and intraneural injection, gave inconclusive results. The same was true as to the alkalinity of the blood, and examination of the urine for acetone and diacetic and lactic acids. A low ammonia content of the urine was found with an extremely low creatin elimination, especially accentuated immediately preceding and during the paralytic attack, with a sudden rise to the normal immediately following the attack. From this the authors conclude that "the symptoms in this case are not solely due to the retention of creatinin or creatin, but that the attacks are due to metabolic disturbance, and that this disturbance may be situated chiefly or primarily, perhaps entirely, in the muscles themselves."

Holtzapple found a diminished excretion of *urea*, and that the worst sufferers were those who showed the most marked diminution in the average daily output of urea. One of his patients showed a very marked increase in the elimination of urinary solids and urea during the attack. Schlesinger in an isolated case found *acetonuria* in the majority of the attacks, and occasionally albuminuria and hyaline casts.

A study of the reported cases suggests a close analogy of the muscular phenomena of the disease and those of myasthenia gravis. In some cases of myasthenia gravis, attacks of paralysis lasting a few days, resembling those of periodic paralysis, are seen.<sup>1</sup>

The rapid exhaustion of the muscular power and the failure of the muscles to respond to the faradic current are suggestive. A careful study of the thymus gland, which is found so frequently diseased in myasthenia gravis, has not been made.

A study of the series reported by Holtzapple bring this whole group of cases into close analogy with periodical ocular paralysis associated with migraine. This series of cases, in which migraine plays such a prominent part and which is not found in any of the other family groups, may be considered to be a connecting link between the periodic family palsies and the periodic oculomotor palsies. In the latter group, a partial or complete oculomotor paralysis of one side, lasting a few days, a week, or longer, is associated with attacks of migraine with a tendency to recur-

<sup>1</sup> Collins. *Vide Jour. Nerv. and Men. Dis.*, 1898, p. 745.

rence at fairly regular intervals. There is a partial or complete oculomotor paralysis affecting one or all the muscles supplied by the third nerve. In cases of this condition which have come to autopsy, a disease process of the third nerve has been found, *i. e.*, in one case a plastic exudate, and in three others tumor formation. Stryzeminiski differentiates a functional and an organic form. Oppenheim agrees with Charcot upon a vasomotor basis for the oculomotor palsy, and considers it to be due either to a vascular cramp giving rise to an ischemic form of paralysis, or a paralysis of the vessel nerves leading to compression by dilatation of the vessels. This would explain the temporary paralysis without damage to the nerve and a terminal degeneration when the condition is frequently repeated.

Holtzapple, following Westphal, explains his cases on the ground of a "vasomotor neurosis affecting the blood supply to the anterior horns which are supplied almost wholly by the anterior spinal artery," and the terminal paralyses "to a slow degeneration of the anterior horns due to frequent disturbance of nutrition and the atrophy of the muscles due to involvement of the trophic cells."

While the association of this one extensive group of cases with migraine points strongly to a central vascular causation, the muscular phenomena noted during the period of paralysis do not support this view. This would bring the disease within the grouping of poliomyelitis, and to a certain extent confirm the opinion of Dana of a recurrent poliomyelitis. Isolated cases in childhood of a flaccid paralysis, with loss of reflexes which last only a few days and then disappear, are not infrequently seen and are usually considered as an abortive or evanescent type of poliomyelitis. The objection to this view and to the other theories of a central nature of the affection is the complete loss of reaction of the muscles during the attack of paralysis to galvanic and faradic stimulation. This would indicate an involvement of the terminal distribution of the nerves in the muscles and the muscle fibres themselves. As a matter of fact, the loss of the myotactic as differentiated from the nerve irritability, together with the changes in the metabolism, point to the muscles rather than to the nervous mechanism. Oppenheim would explain this on the grounds of a peripheral disturbance in the circulation.

**Symptoms.**—The clinical picture is so striking as to be almost dramatic. The patient, as a rule, retires to bed feeling perfectly well, or with slight prodromal symptoms, and awakens during the night with a flaccid motor paralysis which may involve all the voluntary muscles except those of the face, eyes, tongue, the organs of speech and of deglutition. This paralysis may last from a few hours to a few days and disappears gradually or rapidly. In the most severe cases the paralysis is complete and involves the muscles of the trunk and of all four extremities. The muscles of mastication and the tongue muscles may be partially involved. While there is no paralysis of respiration, and while the respiratory rate, as a rule, remains within the normal range, there is often an inability to take a deep breath, and the breathing may be somewhat embarrassed, due to an accumulation of mucus in the throat and bronchial tubes. Bladder and rectal functions are retained.

The *paralysis* is flaccid; the deep and superficial reflexes of the paralyzed muscles are lost. In one of Holtzapple's patients prodromal increase of reflexes was recorded. There are no disturbances of objective sensation. Subjective sensory phenomena are sometimes present; formication, numbness, a sensation of heaviness may precede and continue during the attack. A feeling of soreness of the muscles is not infrequently complained of after the attack passes off.

In Holtzapple's patients, a bulimia commonly preceded the attack, and if the appetite was satisfied, and more particularly by an indulgence in rich or heavy food, an attack was almost certain to follow during the night. Other persons in the same family were not so affected. While prodromal symptoms are not uncommon during the day, the paralytic phenomenon develops, as a rule, during sleep. The affected individual awakens during the night to find himself absolutely helpless as a result of paralysis affecting all four extremities, the trunk, and the neck. Sleep may not, however, be interfered with, the paralysis first manifesting itself on awakening in the morning. In other cases the patient awakens before the attack is fully developed, and many hours may elapse before the paralysis is complete. In some cases the paralysis may develop during the waking hours. In these cases incoördination of the hands and even unconsciousness may usher in an attack. After a period of paralysis varying from a few hours to a week, motor power gradually returns and with it a return of the reflexes and of myotactic irritability.

While there is no paralysis of the bladder or rectum in the strict sense of the term, it is the exception to have urine voided or the bowels moved during the attack. This is probably due to a diminution of secretion and excretion. This is borne out by the examination of the stomach contents in a case of Edsall's. "Even starch digestion was not proceeding and there was a total acidity, showing the whole digestive process to be absolutely at a standstill and the gastric motor power diminished, or abolished for a time." This also explains the lack of desire for food during the attack. During convalescence one or two loose bowel movements or vomiting may occur.

The temperature remains normal throughout the paralytic phenomenon. The pulse may be normal in frequency and volume or may be weak and irregular, with evidence of cardiac dilatation. The electrical reactions have been carefully studied during the attacks and in the intervals, by Oppenheim, Goldflam, and others. In severe attacks there is complete loss of reaction to the faradic and galvanic currents. In the interval between the attacks there may, in some cases, be a diminution in the reaction, a slow vermicular reaction of the muscles, and other evidence of degenerative change. Continued over a long period of time, these muscles may show degenerative atrophy. In most cases in the interval between the attacks the electrical reaction returns to normal.

Besides the severe form of the disease, many cases present a less intense grade both in distribution and the extent of the paralysis. When the paralysis is localized it is most frequently confined to the lower extremities. Paralysis of the upper extremities, the neck, the face, and a hemiplegic form have been described. In some of the cases, and most

frequently the localized forms, there may be only a partial paralysis with slight or marked loss of power. Such attacks may be evanescent, lasting one-half to one hour, or may be prolonged as in the major attacks. In these cases the deep reflexes may be diminished or lost with retention of the superficial skin reflexes. There is likewise a diminution and not a complete loss of reaction to the electrical current.

Abortive and equivalent attacks may occur. In the abortive attack there may be a heavy, sleepy, tired feeling, with slight weakness in the extremities, not sufficient, however, to incapacitate, extending over a period of several days. In some of Holtzapple's series, migraine attacks alternated with the paralytic phenomena and were sometimes associated with it. Consciousness was preserved in all the cases reported, with one exception, in which an attack of unconsciousness ushered in the paralysis.

Between the attacks the patient feels, as a rule, perfectly well. In some cases the patient may awake in the morning completely paralyzed, and be occupied with laborious work by the afternoon of the same day. In other cases the attack passes off more slowly, the full power not being restored in from twenty-four to forty-eight hours. In Burr's case it lasted an entire week.

**Course.**—While cases have been reported as early as the fourth year and as late as the thirty-first year, the majority of the cases develop about the age of puberty. There is a tendency to a diminution in the frequency and intensity of the attacks after middle life. A degenerative type of paralysis affecting both upper and lower extremities and confining the patient to a wheel-chair developed in some of Holtzapple's cases. While this affection is considered by all the other observers as not dangerous to life, sudden death in an attack occurred in six of Holtzapple's cases, giving a mortality of 35 per cent. in his individual group.

**Treatment.**—A study of the family groups above mentioned suggests the idea that the underlying factors vary somewhat in the different groups, and this is borne out by the results of treatment. In two of Mitchell's cases the bromides give no results; in one it had some slight influence. In Holtzapple's cases, 0.5 dram of potassium bromide with 1 or 2 grains of caffeine citrate, repeated in one or two hours, had a decidedly abortive influence when taken in the early paroxysms. In not a single instance did he note the development of an attack when the bromide had been taken during the onset. In Mitchell's cases citrate of potash in doses of 45 to 60 grains a day had "some small but uncertain effect." Administered in the beginning of a seizure in repeated large doses it shortened and mitigated the paralytic period.

Colon lavage, intestinal antiseptics, venesection, hypodermoclysis, and various forms of electrical treatment have been tried with negative results. A study of the factors which determine the attacks and their elimination as far as possible from the life of the individual, together with the maintenance of the physical and nervous tone at the normal level, would appear to be the most rational treatment of a disease which remains with us as it began with Westphal, an unsolved riddle.



## CHAPTER XXI.

### ASTASIA-ABASIA. ADIPOSIS DOLOROSA.

By DANIEL J. MCCARTHY, M.D.

#### ASTASIA-ABASIA.

**Definition.**—A functional disturbance of the neuromuscular mechanism first described by Blocq in 1888 as a morbid state “in which the impossibility of standing erect and walking normally is in contrast with the integrity of sensation, of muscular strength, and of coördination of the movements of the lower extremities.”

**Etiology.**—The disease is not an uncommon complication, and in reality a symptom-group of the functional neuroses—hysteria, neurasthenia, epilepsy, chorea, etc. While it may occur at any age, it is more frequent in youth and early adult life. Knapp's statistics of 50 collected cases show an equal number in men and women. Of these cases, 21 showed hysteria, 3 chorea, 2 epilepsy, and 4 intention psychoses. It may develop spontaneously or during the course of one of the functional neuroses; emotional excitement, traumatism, or exhaustion may be determining factors. It has been noted as a symptom in one case of cerebral tumor localized to the prefrontal area. It is, however, not a symptom of cerebral tumor, and must be considered in this isolated case as an accidental complication.

The absence of any causative pathology even in cases which have existed over a long period of time, the rapid disappearance of the symptoms by suggestion and other methods of treatment, the frequency of its occurrence as the complication of a functional neurosis, place it as a symptom group of purely functional nature. There is a tendency to consider the condition as one of pure hysteria. It may, however, exist without other hysterical stigmata, and in some cases at least, is the manifestation of the extreme fatigue of neurasthenia and negativism of some of the psychoses.

**Symptoms.**—In the simplest form the patient is unable either to stand or to walk and in some cases even to sit (Akathisia), and yet in the recumbent posture retains full power and coördination of all muscles necessary for locomotion. A careful examination in such cases will not show any loss of power, the sensation may be normal, the reflexes show no evidence of organic disease, the bladder and rectal functions are retained, and an examination of the cranial nerves reveals nothing abnormal. It is not at all surprising, however, that in such cases evidence of hysteria in the field of sensation and the special senses should be present. When the condition develops upon a basis of neurasthenia, marked fatigue and quickened reflexes with paresthesia are to be expected.

Apart from the typical and complete case, there is a large group of cases in which the ability to stand and to walk is not entirely lost, but interfered with to a greater or less degree, as is shown by a sudden or general loss of power with incoördination. This is of an irregular type, and can be easily distinguished from the ataxic gait of the tabetic or that seen in cerebellar disease. Even in cases which show a complete loss of the ability to stand or walk the patients are still able to walk on all-fours, to walk backward, and in some cases to swim (Oppenheim).

The fatigue consequent upon muscular exertion, seen in neurasthenia, may, in some cases, be so intense as to constitute the basis for the loss of power, accentuated and to a certain extent determined by the nervous excitement and lowered will-power. In other cases the disease has its origin in a psychasthenia. In an individual who has a dominating fear of paralysis, after prolonged introspection an astasia-abasia may develop upon a moderate or marked grade of fatigue. Excessive fatigue, more particularly when associated with mental stress, may be the starting point in one who is neither a psychasthenic nor an hysterical patient. In the nervous or timid child, fear of pain after a fracture or other injury to the foot may cause the condition.

Ziehen gives the following classification:

1. Hysterical astasia-abasia dependent upon the subconscious idea implying an inability of the patient to stand or walk. He places this in the group of hysterical palsies.

2. Hypochondriacal astasia-abasia dependent upon the conscious but false idea of the inability of the patient to stand or walk. This may arise as a primary delusion or be dependent upon paresthesia, visceral disturbance, diplopia, vertigo, cardiac palpitation, etc.

3. Affective or emotional astasia-abasia caused by a subemotional shock. An attempt to walk is associated with a sudden overwhelming fear, inhibiting the motor function.

4. The psychasthenic form of astasia-abasia dependent upon a morbid impulse or concept that develops on a false idea of inability to walk or stand, or paralysis, or a fear of falling. This form is dependent on defective will power.

**Diagnosis.**—Astasia-abasia must be differentiated from paralysis due to organic disease of the spinal cord, cerebellar disease, labyrinthine vertigo, and myasthenia gravis. The absence of ankle clonus or Babinski reflex, or bladder and rectal disturbance, and of sensory disturbance of organic distribution, will easily exclude organic disease of the cord. In cerebellar tumors, optic neuritis and other evidence of intracranial pressure, together with the typical ataxia, will be present. The rapid muscular fatigue seen in *myasthenia gravis* after exertion may be easily mistaken for a partial form of astasia-abasia. This is particularly true when the symptoms of the onset are most marked in the lower extremities. The loss of power in these cases is as marked, after repeated muscular exertion, when the patient is recumbent as when he is standing. The myasthenic reaction will establish the diagnosis.

*Akinesia algera*, a condition based upon the same neurotic basis as astasia-abasia, should be differentiated from it. In this, all the voluntary

movements are associated with intense pain, which increases in severity if the movements are continued. The pain may become so intense as to affect the entire body upon the slightest motion, and in this way produce a pseudoparalysis. The absence of pain in simple and uncomplicated astasia-abasia should differentiate it from the above affection.

**Prognosis.**—This is, as a rule, favorable. In most cases the condition yields easily to treatment. Some cases are very resistant and prolonged. The prognosis may be said to be dependent upon the underlying cause and the ease with which it may be controlled or removed. In cases which are permitted to run for a long period of time, contractures may develop which may necessitate vigorous and even surgical treatment. This complication is likely to develop when the astasia-abasia is associated with a hypertension of the muscles.

**Treatment.**—This does not differ essentially from that detailed under the functional neuroses. Removal of any causative factor which may be present, the reestablishment of confidence, the training of the will power, and reeducation as to the method of walking, standing, etc. (following the general plan outlined by Frenkel in tabes), are indicated. In conditions of lowered nerve tone, partial or complete rest treatment, with or without hydrotherapy, is advisable. Suggestion by means of static electricity, and in stubborn cases under hypnotism, may be necessary. It should be remembered that the cure of astasia-abasia in a patient with other symptoms of hysteria or neurasthenia is simply the relief of a symptom-group, and not the cure of the disease.

### ADIPOSIS DOLOROSA (DERCUM'S DISEASE).

This was first described by Dercum<sup>1</sup> in 1888, from a case studied in the wards of the Philadelphia Hospital (Blockley).

**Etiology.**—It is a disease preëminently of middle life. Cases, however, have been described as early as the eleventh year and as late as the seventy-eighth year, but these are exceptions. The vast majority of cases are found in the female sex. A few cases have been reported in men, one of these by Ewald,<sup>2</sup> in the forty-seventh year. A history of alcoholic excess is not infrequent. Traumatism and emotional excitement have also been given as causative factors. Syphilis as a factor has not been given the consideration which its frequency deserves. The disease develops, as a rule, upon a neuropathic basis, and several patients have terminated their existence in insane asylums. While heredity is not an important factor, in Cheever's<sup>3</sup> case the father and sister were also affected. Hammond reports two cases occurring in sisters. The majority of cases in women have developed at the time of the menopause or shortly afterward. One case followed an abortion, and in another pregnancy was given as the starting point of the disease.

<sup>1</sup> *University Medical Magazine*, December, 1888. See also *Am. Jour. Med. Sc.*, November, 1892, and *Jour. Nerv. and Ment. Dis.*, August, 1900.

<sup>2</sup> *Berl. klin. Woch.*, 1895, Nos. 2 and 3.

<sup>3</sup> *Brit. Med. Jour.*, 1904, i, 781.

**Pathology.**—To understand the position of adiposis dolorosa as a separate affection, it will be necessary to make a rapid survey of the other affections of a fatty nature. Apart from simple obesity, the following forms have been described: 1. Adiposis tuberosa simplex of Anders.<sup>1</sup> 2. Adiposis cerebialis.<sup>2</sup> 3. Adenolipomatosis.<sup>3</sup> 4. Multiple lipomatosis.

1. **Adiposis Tuberosa Simplex.**—This affection, at least in some of the cases, resembles adiposis dolorosa very closely. In patients who present for the most part the condition of simple obesity, Anders found localized fat tumors, sometimes painful to pressure, scattered through the abdominal fat. These differ, however, both in their formation and in their disappearance under simple dietetic and hygienic measures, from the nodules seen in adiposis dolorosa.

2. **Adiposis Cerebialis.**—This condition, described by Fröhlich as dystrophia adiposo-genitalis, has a very close analogy to adiposis dolorosa. In its simplest form it consists of an excessive general adiposis developing during the course of a brain tumor, which in most cases has been found at autopsy to involve the pituitary body, but which in one case was found to involve the pineal gland (Marburg). A close association has been found in this group with defective genital development.

3. **Symmetrical Adenolipomatosis.**—This condition, first described by MacCormac, consists of large fat masses localized to the region of the neck, the axilla, and the trunk, and associated with asthenia, mental irritability, apathy, hypochondria, enlargement of the spleen, acceleration of the pulse, and decrease of small mononuclear cells in the blood. These fat masses, even when of great size, may be associated with general emaciation. Microscopic examinations of excised portions have shown the presence of large and sclerosed lymphatic glands in the fatty tissue. An increase and diminution in the size of the fat during its development led Launois and Bensaude<sup>4</sup> to consider the disease to be of lymphatic origin.

4. **Multiple Lipomatosis.**—Multiple isolated lipomatous tumors have occurred symmetrically placed in different portions of the body. On account of the symmetrical arrangement and the association of the tumors with tabes, general paralysis, sciatica, etc., it has been assumed that the nervous system and more particularly the trophic centres in the spinal cord were the cause of the affection. There is, however, practically no evidence to support such a theory. As many as two thousand of these lipomas have occurred in the same individual.

Careful study of these various forms of diseases of the fat tissue will show a certain similarity. A study of the records of seven cases of adiposis dolorosa that have come to autopsy has shown lesions in the thyroid gland in the nature of atrophy with compensatory hypertrophy in all cases. Tumors of the pituitary have been found in three cases, an interstitial neuritis in the fat tissue in two cases, and a slight sclerosis of the posterior columns of the cord in one case. In one of these cases

<sup>1</sup> *Am. Jour. Med. Sc.*, 1908, cxxxv, 325.

<sup>2</sup> Marburg, *Wien, med. Woch.*, 1908, No. 49.

<sup>3</sup> MacCormac, *St. Thomas' Hospital Reports*, 1883, xiii, 287.

<sup>4</sup> *Nouvelle Iconographie de la Salpêtrière*, 1900, xiii, 41, 184, 243.

(Dercum and McCarthy), presenting a typical clinical picture of adiposis dolorosa, a tumor of the pituitary was associated with a hypoplasia of the genital organs, extensive lymphoid infiltration of the fat and hemolymph glands. This case forms a connecting link between adiposis dolorosa, adiposis cerebri, and symmetrical adenolipomatosis. A case of adiposis, presenting clinically the picture of adenolipomatosis, in the writer's ward at the Philadelphia Hospital was found at autopsy to be a case of psammocarcinoma of the thyroid, with extensive metastasis in the fat tissue in the neck, destruction of the posterior portion of the pituitary body by an extensive multilocular cyst, sclerosis and old hemorrhagic infiltration of the adrenals, and extensive lymphadenitis with lymphoid infiltration in the fat masses, thyroid, pituitary, and adrenals.

The compensatory action of the thyroid and pituitary bodies has been demonstrated experimentally. It would appear from the frequency of the lesions that the thyroid was mainly at fault in the production of the disease of the fat tissue. In the case of adiposis cerebri, the lesions of the pituitary and the improvement after the removal of the pituitary tumors would point to the pituitary as the source of the trouble.

The influence of the *lymphatic* system must be of importance. It has been found diseased in the cases above described, and in the tumor masses excised from cases of adenolipomatosis. Chipault and others have shown that localized lymph stasis may be transformed into fat tissue by a conversion of the connective tissue into fat cells. Whether the thyroid and the pituitary produce the changes above described, by disturbance of metabolism through some perturbation of the internal secretion, or whether the changes noted are secondary to changes in the lymphatic system, is a subject for investigation.

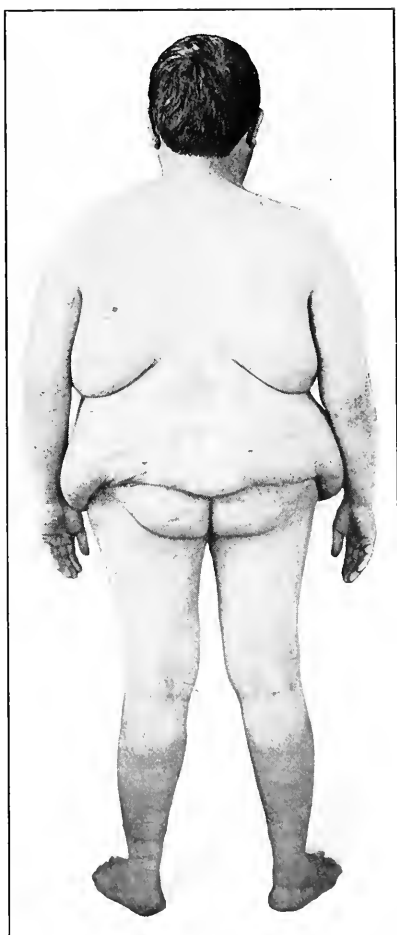
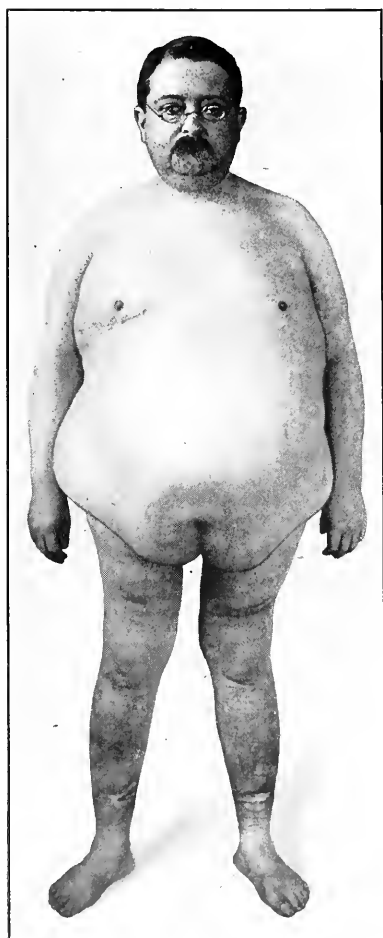
The cause of the pain and tenderness in the fat masses in adiposis dolorosa is an interesting subject for speculation. Quite a number of the cases of adiposis dolorosa have shown some organic change in the central or peripheral nervous system. Tenderness over the nerve trunks, sensory changes, atrophy and degeneration of the muscles have been noted. Traumatism and syphilis have been noted as causative factors. Such factors acting upon the peripheral nervous system concerned with the nutritive changes in excessive adipose formation would easily produce low-grade interstitial changes in the nerves of the adipose tissue, and thereby cause the pain and tenderness. The pathological lesions of urticaria factitia described by Gilchrist present a pathological picture of transient acute inflammation. A similar lesion continued over a longer period of time and frequently repeated could easily cause the nerve changes and symptoms above noted. Such changes could more easily be produced in nerves whose function and nutritive tone were lowered by some metabolic or exogenous intoxication.

**Symptoms.**—The disease begins, as a rule, in a slow and insidious manner, in the formation of either excessive deposits of fat scattered diffusely over the trunk and extremities, or in the form of irregular deposits of fatty tumors. It may develop as an independent affection in which the adiposis constitutes the disease, or may, on the other hand,

develop in one who is already of a fatty habit. The development of the disease is best studied in the latter group. A woman in middle life, of moderate obesity, develops, with or without prodromal symptoms, areas of painful swelling, irregular in size and distribution, somewhat elevated above the surrounding fat, the overlying skin of a normal appearance, slightly reddened, more often congested, and rarely with distended veins. These areas vary from one to two or four inches in diameter, are soft and somewhat oedematous to touch, very painful to palpation, and very often associated with a burning or lancinating subjective sensation of pain. After several days this condition disappears, leaving an indurated area in the fat tissue. This area from time to time is subject to a recurrence of the same condition less intensified, until finally a distinct nodular tumor formation of considerable resistance and consistence is left. This nodule is sensitive to pressure, at times even to light palpation, and constitutes the basis of the disease, *adiposis dolorosa*. Similar areas develop elsewhere through the fat tissue of the trunk and extremities, never affecting the face, the hands, or the feet. With further development there may or may not be an excessive deposit of fat. In the latter event the nodules may be seen distinctly elevated above the surrounding tissue, having somewhat the appearance of a multiple lipomatosis. In diffuse cases the fat is deposited in large masses, localized to certain areas, such as the abdomen, chest, thighs, etc., or massed in a general diffuse way over the entire trunk. Examination shows indurated areas, sensitive to palpation and pressure, scattered here and there throughout the fat masses. In the acute stage the swollen mass often reminds one of "caking breast." As the disease progresses to a full development, the areas of acute swelling and tenderness become less frequent, and finally disappear altogether. Nodules in the fat remain as circumscribed masses, sensitive to pressure, and associated with diffuse pain of varying character described as neuralgic, rheumatic, etc. In some cases there is no history of the acute swollen areas, but, on examination, fully developed, painful, tender masses are present. In some of the diffuse cases, folds of fat give a sensation to the touch as of a mass of worms. In some ill-defined cases, not properly classed with this disease, the whole fat tissue is sensitive without the formation of nodular areas and without spontaneous pain.

*Pain* is a very prominent symptom present in all well-marked cases at some time or other, and may precede the development of the fatty nodules. It is often the only symptom complained of, and may be a sharp lancinating pain or a dull ache, and is often described by the patient as a burning pain. It does not follow the course of the nerves, and is often associated with tenderness along the nerve trunks. While the painful and tender fatty areas constitute the main and the essential symptoms, a rather wide group of associated symptoms are frequently present. The most common of these is *asthenia*. It is exceptional to find a well-developed case of *adiposis dolorosa* in which this symptom, in some grade, is missing. It may be nothing more than the nervous apathy and sluggishness of very fat people, but in the majority of cases is so marked as to be considered of pathological origin. Psychic symptoms are not infrequent.

PLATE XI



Adiposis Dolorosa. (Case of Dercum and McCarthy.)

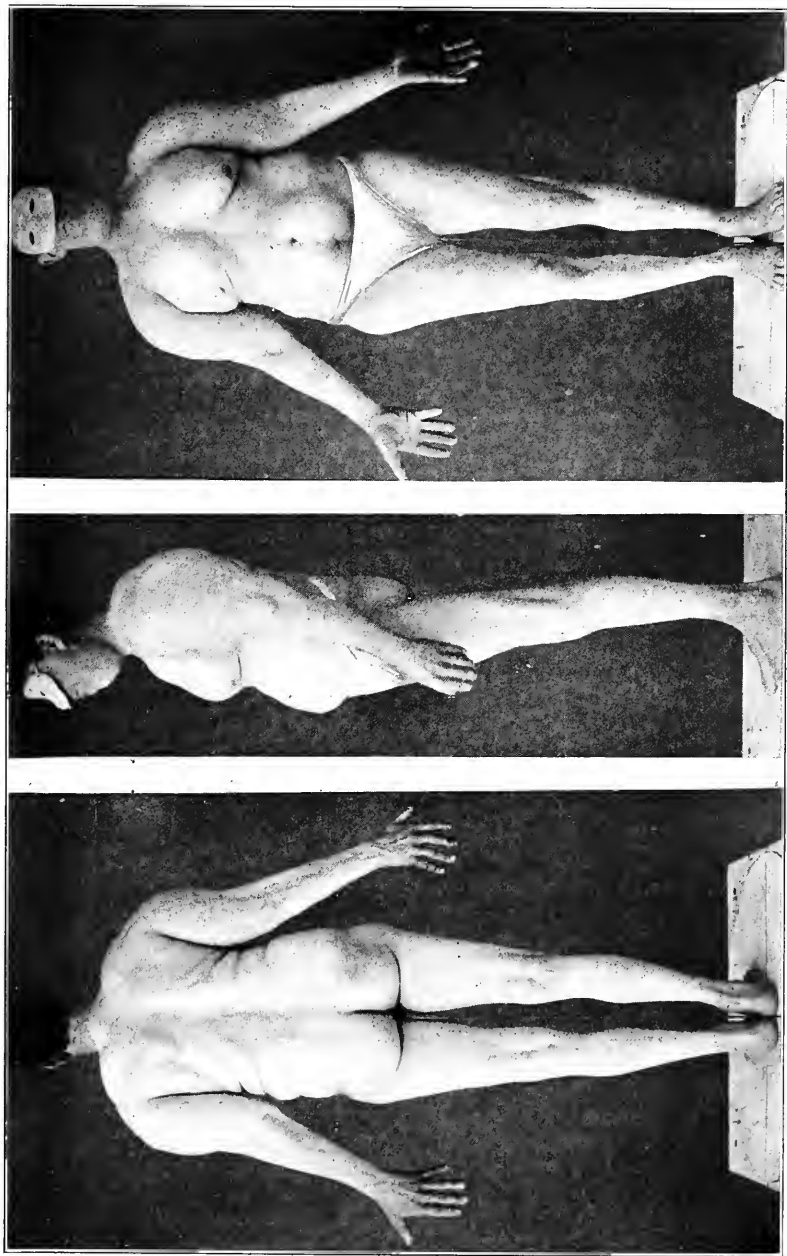
Tumor of the pituitary body; thyroïdal changes; defective development of the testicles.

From the *American Journal of the Medical Sciences*, December, 1902.





PLATE XII



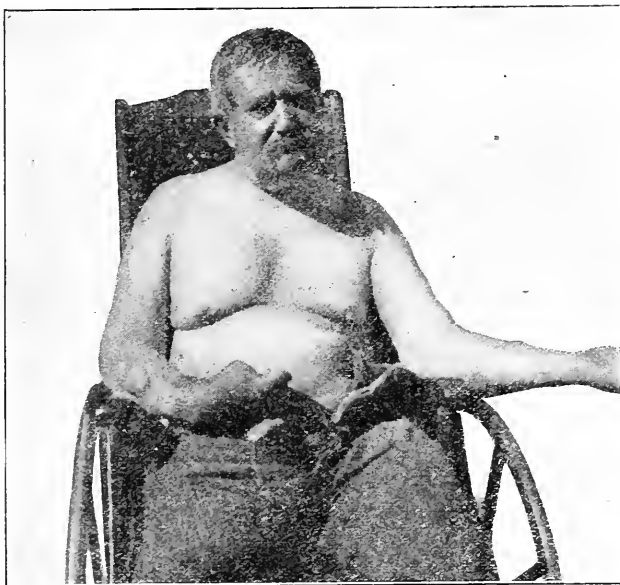
Adenolipomatosis. (Case of Dr. C. K. Mills.)



The most common of these is a querulous irritability. Mental apathy and psychic depression are most common in the asthenic cases. Delusional states and dementia sometimes necessitate the incarceration of the patient in an asylum. The reflexes are diminished and sometimes lost, but may be increased.

In Dercum's first case there were areas of anesthesia and hyperesthesia, and several cases showing sensory changes have since been reported. Disturbance of the vasomotor system has been frequently noted. Variations in sweat secretion, cyanosis of the extremities, dermatographia, trophic ulcers, and subcutaneous ecchymoses have been described.

FIG. 47



Multiple symmetrical lipomatosis. Hemiplegia.

**Prognosis and Treatment.**—The prognosis as to life is good, but as to cure is bad. In the majority of cases the disease is very refractory. Thyroid gland in ascending doses has been of benefit, in a small number of cases, in reducing the pain and the fat tumors. Aspirin and salicylates have been used with some benefit to relieve the pain and tenderness. Massage, baths, diet, etc., have been of only temporary benefit. The tenderness over the nerve trunks has a tendency to disappear as the disease advances.

## CHAPTER XXII.

### SYPHILITIC DISEASES OF THE CENTRAL NERVOUS SYSTEM.

By B. SACHS, M.D.

THE discovery of the *Spirochæta pallida* (Schaudinn) and the sero-diagnostic investigations of Wassermann and his followers have brought about a great advance in the recognition of syphilitic diseases of the nervous system. It is impossible to over-estimate the support which these methods give to the accuracy of diagnosis. On the other hand, laboratory methods are an addition to, and not a substitute for, clinical observations. A positive reaction of the cerebrospinal fluid gives certain evidence that the central nervous system is the seat of specific disturbance. The cerebrospinal content, the increase in the number of lymphocytes, the excess of globulin, make the syphilitic nature of cerebrospinal disease still more positive. While the absence of all these changes in the cerebrospinal fluid may not militate altogether against the diagnosis of syphilis of the nervous system, our present-day understanding of the subject makes it at least improbable that there is any very characteristic specific disease present.

At the time of the publication of the first edition of this treatise neurologists were still in the habit of making a very marked distinction between syphilitic and parasyphilitic diseases of the central nervous system. *Parasyphilis* was regarded as a form of degeneration following upon syphilis but not actually due to the virus of the disease. The discovery by Noguchi of the presence of the spirochæta pallida in the cortical tissue of patients dying of dementia paralytica has proved that parasyphilitic diseases, so-called, are essentially syphilitic, though there may be certain differences in kind and degree in the case of late syphilitic manifestations. MacIntosh and Fildes<sup>1</sup> hold that these are not "Syphilitic lesions of an unusual or particular form, but that they resemble in all essentials such lesions found elsewhere in the body." We shall do well, therefore, for the purpose of this article, to maintain the clinical forms formerly established, but the student is to remember that, however different the clinical manifestations may be, all the forms herein discussed are etiologically of equal value. They are all the expression of a luetic infection which may differ in intensity or distribution, but in every instance the disease is considered to be due to an infection caused by the spirochæta pallida and the changes wrought in the central nervous system are a result.

The British authors quoted above have been at special pains to prove that the primary sores, the cutaneous lesions of the secondary period and the tertiary lesion, the gumma, revealed the same histological character-

<sup>1</sup> *Brain*, 1914, xxxvii, 141.

istics, viz., perivascular collections of lymphocytes and plasma cells associated with a proliferation of the cells of the affected vessels. All these lesions contained the *Spirochæta pallida* and this organism represents the irritant about which the cell reaction is found. There is a qualitative difference, however, and in the tertiary process, for instance, the spirochætes are present in a very much smaller number than they are in the primary or secondary form.

According to the more recent view, it is well to regard the entire syphilitic process as passing through an acute and chronic stage. In the acute stage the spirochæte may wander into any of the internal organs and a varying degree of degeneration of the parenchymatous tissue may follow. As the acute stage of syphilis subsides there is a general destruction of the spirochætes, but some may be retained somewhere in the system or in some organs, so that isolated foci may be maintained for some considerable period of time, possibly years. The organisms lie dormant but are capable of revivification and may thus give rise to the very late tertiary lesions. If the lesion is an interstitial one there may be very few symptoms developed, whereas if the parenchymatous tissue is involved the cellular tissue will degenerate and the special function of the organ is certain to be affected. It follows from this that in interstitial lesions the effect of treatment will be much more apparent than in purely parenchymatous lesions.

It has been well known for many years that, like other poisons, the syphilitic virus has distinct organs and sites of predilection. Thus, the liver and the testicles have been known to be more often affected than the spleen or the lungs. It was not appreciated, however, until lumbar puncture became a routine practice in many of our hospital services that the central nervous system is involved in the disease process with astonishing frequency. Ravaut has shown that meningeal irritation was evidenced in 71 out of 118 cerebrospinal fluids examined even when there were no obvious nervous manifestations of central syphilis. The headache, the drowsiness, and the mental derangement which have occasionally been observed in the early stages of syphilitic infection must now be attributed to the early involvement of the central nervous system. Within my own experience distinct increase of lymphocytes in cerebrospinal fluid has been found within three weeks of the initial lesion.

Bearing in mind the fact that all these forms have a common etiology, with variations as to intensity and distribution, it is, nevertheless, practical to differentiate between the following types of syphilitic disease: (1) Cerebrospinal syphilis; (2) acute and chronic specific myelitis; (3) syphilitic spinal paralysis; (4) tabes dorsalis; (5) general paresis.

**Cerebrospinal Syphilis.**—Almost every manifestation of brain and spinal cord disease might be enumerated under this heading. At least, there are very few conditions pointing to focal disease in the brain or in the medulla spinalis which might not at one time or another be taken to be due to preceding syphilitic infection. It is not the intention to enumerate all such diseases, but to select for especial mention a few of the clinical conditions which are so distinctly specific that they can by clinical examination alone be recognized as a form of syphilis of the nervous

system. More than twenty-four years ago the writer<sup>1</sup> attempted to impress this fact upon the medical public by helping to establish a condition known as multiple cerebrospinal syphilis. The resemblance in name to multiple cerebrospinal sclerosis was favored because, as a matter of fact, the differential diagnosis between these two conditions must frequently be made.

By *multiple cerebrospinal syphilis* we mean an affection which involves distant parts of the cerebrospinal axis and gives rise to a multiplicity of symptoms, some of them due to a lesion of parts as widely separated as are the brain axis and the sacral segments of the cord.

**Etiology.**—Under this heading nothing need be said except that this form is frequently due to a more or less recent infection and may appear at any time within a few months to a few years after the initial lesion. Nowadays the strongest possible corroboration of a suspected diagnosis may be had from the examination of the cerebrospinal fluid. Nonne<sup>2</sup> claimed at first that in these very cases the Wassermann reaction of the cerebrospinal fluid was often negative; but the more improved methods of recent years have shown that the Wassermann reaction of the cerebrospinal fluid is positive in almost all the cases (Kaplan claims 100 per cent.), and that the globulin content is excessive in a very large percentage of the cases. At the time of a first examination, the amount and character of preceding treatment must be taken into account. But a negative reaction cannot be taken to prove the absence of syphilitic poison.

This form of specific disease may safely be said to be due in a considerable number of the cases to a chronic syphilitic involvement of the *meninges* of the brain and spinal cord. According to the predominance of the symptoms we may, therefore, have a pachymeningitis or leptomeningitis cerebri specifica, or a pachymeningitis or leptomeningitis specifica spinalis. That from this meningitis the process may extend into the brain or spinal cord need not be insisted upon, and if some writers speak of meningo-encephalitis specifica or of meningomyelitis specifica, the morbid process itself unquestionably remains the same. The early and persistent headaches are the clinical expression of a chronic convexity meningitis of luetic origin. With this widespread involvement of the membranes of the brain and spinal cord there may be special gummatous deposits, and this accounts for the fact that in so many instances we have symptoms pointing with special force to involvement of remote parts. A gumma in the vicinity of the crus may be associated with a general leptomeningitis, and we can well understand, therefore, that in such a case there may be complete ophthalmoplegia interna and externa, without other cranial nerve symptoms, associated with a spastic paraplegia of the lower extremities and involvement of vesical and rectal functions, since the dorsal and lumbar portions of the cord are sites of predilection for the specific spinal pachymeningitis and leptomeningitis. Aside from involvement of the brain and spinal cord substance and the meninges, we may have a specific endarteritis as the sole expression of a

<sup>1</sup> *New York Med. Jour.*, 1891, liv, 309.

<sup>2</sup> *Syphilis u. Nervensystem*, second edition, Berlin, 1909 (contains full literature to date).

more or less chronic syphilitic process. In such conditions attacks of hemiplegia due to thrombosis are very apt to occur, and there is no doubt that specific disease of the spinal cord bloodvessels with subsequent softening may give rise to various clinical groupings pointing to involvement of the spinal cord itself.

It is characteristic of syphilitic disease of the nervous system that the granulation tissue (granular proliferation) starts from the smallest capillaries, either from the capillaries of the connective tissue or from the capillaries (*vasa nutritia*) of the larger bloodvessels. Oppenheim was one of the first to lay special stress upon the gelatinous character of the specific thickening of the spinal and cerebral meninges. Mention must also be made of the fact that a primary degeneration of the cranial nerve nuclei may be an expression of a general syphilitic infection.

**Symptoms.**—The disease may come on in acute or subacute fashion. Headaches and vomiting are among the earliest symptoms. These are sufficient, in many instances, to arouse the suspicion of brain tumor; if, as happens often enough, optic neuritis is also present and rapidly progressive, the diagnosis of brain tumor may appear to be sufficiently well founded to justify cerebral decompression. It will be well, therefore, to make the specific tests of blood and cerebrospinal fluid before suggesting decompression.<sup>1</sup> The onset of the localizing symptoms may be preceded by general symptoms such as are known to be due to a general specific disease, particularly of the brain, and it may be noted that in a very large majority of the cases cerebral symptoms precede those of a spinal character. The earliest symptoms of this condition are apt to point to involvement of the base of the brain and particularly to involvement of the various cranial nerves, and among these the third nerve and the sixth are most frequently affected, while in others the fifth, seventh, eighth and twelfth nerves may be involved, according to the special distribution of the lesion. An early ptosis, an ophthalmoplegia externa and interna, unilateral deafness, abnormalities of sensation and even dissociation of sensation within the distribution of the fifth nerve, paralysis of one or both vocal cords, paralysis and atrophy of the tongue with accompanying disturbance of deglutition and of speech, and, of course, unilateral facial paralysis—all these symptoms may appear early in the course of a multiple cerebrospinal lues. Moreover, cranial nerve symptoms may be associated with paralysis of the upper and lower extremities. Not infrequently the upper extremities escape, so that we have the peculiar association of cranial nerve palsies with a paraplegia of the lower extremities. In the vast majority of the cases this paraplegia, whether of the upper or lower extremities, is of the spasmodic order, due, no doubt, to the more frequent involvement of the lateral portions of the cord, or to the fact that when there is a meningeal infiltration the invasion from the meninges affect first the tracts running through and near the lateral columns. That the spasmodic paraplegia would be associated with increase of the deep reflexes and with a disappearance of the superficial reflexes in the regions affected goes without saying. The vesical and rectal functions may or

<sup>1</sup> During the past years I have had decompression done in well established forms of luetic optic neuritis to prevent further injury to the optic nerves.

may not be disturbed. The paraplegia may at times be flaccid, and Hoffman<sup>1</sup> has reported cases on the order of poliomyelitis anterior due to syphilis, while Spiller<sup>2</sup> has reported a syphilitic acute anterior poliomyelitis due to thrombosis of the cervical anterior median spinal artery.

The clinical picture may, therefore, involve one or more parts of the central nervous system, and these parts may be remote from one another, so that the mere fact that a single lesion cannot explain all the symptoms in a given case, and that there must be a multiplicity of lesions, points with strong probability to a general luetic process.

In the article on *tabes* reference is made to the *pseudotabes syphilitica*. We need not, therefore, at present refer to the possible involvement of the posterior columns of the cord giving rise to symptoms resembling *tabes*, and to their association with symptoms pointing to distinct involvement of the brain axis. But cases of this sort can be differentiated from true *tabes* by the very multiplicity of symptoms, by the association of tabic symptoms with a number of ocular nerve palsies, and not merely by the association with ptosis or with rectus externus palsy, as happens in some cases of *tabes dorsalis*. Whatever the localization may be that is indicated by the clinical symptoms, there are other peculiarities which lend support to the diagnosis of a multiple cerebrospinal lues. The writer summarized these so long ago as 1893.<sup>3</sup> They are: (1) The unusual distribution of the disease over the greater portion of the brain and spinal cord axis; (2) the relatively slight intensity of the morbid process as compared with the extensive area involved, evidenced by the preservation of some of the functions with complete loss of others; (3) a rapid disappearance of some of the symptoms and a very chronic persistence of others; and (4) the frequent history of other cerebrospinal accidents. Thus, some of the patients give a history of a preceding hemiplegic attack with more or less complete recovery. Others give a history of temporary paralyses, of temporary aphasia; still others, of a preceding attack of paraplegia which may have lasted a few weeks or a few months, followed by satisfactory recovery. It is the remissions and exacerbations, the recoveries and relapses, that are more characteristic of a cerebrospinal syphilitic process than of any other condition.

No investigation is complete without a thorough examination of the cerebrospinal fluid. In hospital practice it may be feasible to do lumbar puncture in every form of brain or spinal syphilis; in private practice it may be well to limit the practice strictly to those cases in which the diagnosis is somewhat in doubt, or in which it is desirable to gauge or determine the effects of treatment by repeated examination of the fluid. In several of my own patients, the temporary relief of symptoms, effected by the withdrawal of fluid has been so marked, that the patients have requested repeated lumbar punctures.

The facts to be noted are as follows: A positive Wassermann reaction of the blood indicates constitutional lues (acquired or hereditary). It does not, however, prove that the disease itself is luetic. A negative

<sup>1</sup> *Neurol. Centralbl.*, October 16, 1909.

<sup>2</sup> *Jour. Nerv. and Ment. Dis.*, 1909, xxxvi, 601.

<sup>3</sup> Syphilis of the Spinal Cord, *Brain*, 1893, xvi, 405.



reaction of the blood does not exclude syphilis, particularly if the blood has been taken from a patient who has been subjected to much anti-syphilitic treatment. In general paresis the Wassermann reaction is so rarely negative, that negative findings may militate against the diagnosis of that disease.

**Diagnosis.**—The chief task is to differentiate multiple cerebrospinal syphilis from *multiple cerebrospinal sclerosis*, which can be distinguished from the latter by the absence of intention tremor, of nystagmus, and of scanning speech, and, above all, by the frequent remissions in the course. And yet remissions are characteristic also of disseminated sclerosis. However, in disseminated sclerosis there is rarely such complete recovery as one not infrequently sees in multiple cerebrospinal syphilis. At the present day the examination of the cerebrospinal fluid and of the blood will give valuable assistance, and if the findings in the blood and in the spinal fluid are positive, disseminated sclerosis may be excluded, since it is tolerably certain that it is the rarest thing in the world to have disseminated sclerosis develop on the basis of a preceding syphilitic disease.

The only other widespread affections of the central nervous axis which might resemble multiple cerebrospinal syphilis are a *cerebrospinal tuberculosis* and a general *carcinomatosis* or *sarcomatosis*. From tuberculous disease the syphilitic affection may be distinguished by the more rapid course of the former and by the fact that the tuberculous trouble very often leads to a strictly limited basilar or spinal meningitis with formation of one or more solitary tubercles. Moreover, the tuberculous process affects the lower portion of the brain axis and the upper portion of the spinal cord much more frequently than it does the dorsolumbar region. A general sarcomatosis or carcinomatosis may give rise to symptoms somewhat resembling those of multiple cerebrospinal syphilis, but, as a matter of fact, a difficulty in diagnosis rarely arises, for the preceding history of primary neoplasm in some other organ, particularly in the breast or in some of the glands or bones, puts the physician on his guard. The distribution of the symptoms may be very similar, but the greater gravity of the disease is easily recognized. As a rule, the symptoms are steadily progressive, although, as in a patient recently seen, the symptoms may be stationary over a period covering six to eight months. The autopsy showed a general carcinomatosis extending all the way up and down from the cervical to the sacral region, involving the vertebræ and pressing upon the spinal coverings, but the spinal cord itself had only been affected by compression. In other cases, however, the carcinomatous process may cause complete destruction of the cord. To quote from a previous article of the present writer, we may state "that there is a frequent combination of symptoms that can be recognized as a special form of disease—a multiple cerebrospinal syphilis due chiefly to a specific meningitis of the brain and cord, which is apt to form special deposits in different parts of the central nervous system, or by proliferation to make inroads into the brain or cord, and that the association of cranial nerve affections with special symptoms, most frequently of a spastic type, in the presence of a distinct history of syphilitic contagion and of repeated remissions, gives strong evidence of this special form of disease."

**Acute and Chronic Syphilitic Myelitis.**—The frequent localization of the syphilitic process in the dorsal and lumbar regions of the spinal cord gives rise to a clinical picture pointing to an acute, subacute, or chronic myelitis. The absence of root symptoms (pains, paresthesia, etc.) indicates that the spinal meninges have escaped, but the association with distinct cerebral luetic symptoms, such as pupillary immobility or strabismus, goes to show that the specific myelitis is an especially restricted form of the cerebrospinal syphilitic disease. In the variability and incompleteness of the symptoms we have the same earmarks of the luetic process that were dilated upon in the section on cerebrospinal syphilis. The features of syphilitic myelitis are discussed elsewhere (page 237), and require no extended discussion here. According to the special areas of the cross-section of the cord which may be involved in the luetic process the symptoms will naturally vary. The clinical pictures of acute or chronic poliomyelitis anterior or of amyotrophic lateral sclerosis may be presented when the disease is, in reality, an acute or subacute incomplete luetic myelitis. Almost any form of myelitis may occur due to syphilis. In making the diagnosis, it is of the utmost importance to consider other evidence of constitutional syphilitic disease which the patient may have presented at the time of examination or in preceding years. The writer has seen patients treated at one time for a transitory ocular palsy, at other times for an attack of hemiplegia, and then again for a spastic paraplegia of the lower extremities. One patient, who has passed through these three phases, in addition has developed a condition of specific dementia bearing a close resemblance to general paresis. The previous occurrence of such accidents, the frequent recoveries from more or less serious paralytic conditions, are features common only to syphilitic spinal disease. It should also be remembered that a specific meningitis or meningomyelitis may go hand in hand with the formation of one or more gummata, and the difficulties of diagnosis may therefore be increased by the fact that symptoms pointing to tumor of the cord may either precede those of myelitis or be superadded to them.

Under *normal* conditions, the cerebrospinal fluid is under pressure of 90 to 130 mm. (Nonne); it contains few, if any, cells; surely not more than 5 or 6 per mm. (Fuchs, Rosenthal, Chamber). The Wassermann reaction is negative (whether 0.2 cm. or larger quantities up to 1 cm. of the fluid be used). The globulin reaction is negative.<sup>1</sup>

Under *pathological* conditions, the cerebrospinal fluid flows out under increased pressure. The globulin content is markedly increased; there is a marked lymphocytosis; all these symptoms have corroborative value, but may occur in any organic infectious, irritative, luetic or non-luetic disease. A positive Wassermann reaction of the fluid clinches the argument in favor of syphilis of the central nervous system (to avoid possible error quantities of fluid from 0.2 to 1 cm. should be employed).

**Specific Spinal Paralysis (Erb's Type).**—This is discussed elsewhere (page 246) and is included here for completeness. Erb called attention to a series of symptoms which he considered pointed indubitably to

<sup>1</sup> For all serological details consult Kaplan's *Serology of Mental and Nervous Diseases*, Philadelphia, 1914.

syphilitic spinal paralysis, but Oppenheim and the writer, while recognizing the frequent occurrence of Erb's type, were more inclined to consider it *a* syphilitic spinal paralysis and not *the* syphilitic spinal paralysis. The experience of later years has strengthened this view. Erb's type was characterized (1) by the usual symptoms of spastic paraplegia with its peculiar gait, carriage, and movements; (2) by marked exaggeration of the deep reflexes; then by muscular contractures which are slight as compared with the exaggeration of the reflexes. Involvement of the bladder, a slight, yet distinct, disturbance of sensation, gradual onset of the disease, and a decided tendency to improvement, were further characteristics. Erb inclined to attribute this special form to a diffuse meningomyelitis, but Nonne has shown very conclusively in several cases which came to autopsy that Erb's type of syphilitic spinal paralysis may be the expression of a combined systemic degeneration involving the pyramidal and cerebellar tracts and the tracts of Goll.

Before concluding, a quotation is taken from the chapter in the writer's book on the *Nervous Diseases of Children*: "One may suspect specific disease of the cord if the patient presents symptoms of paralysis, whether they be of the spastic or flaccid character and whether the contractures be slight or not, provided the patient furnishes evidence of a morbid process affecting a very large part of the cord and yet showing a relatively slight intensity at any given level of the cord. He may, for instance, exhibit the symptoms of extreme paralysis, spastic or atrophic, with partial or slight anesthesia of the parts paralyzed, with little or no involvement of the bladder, or, as often happens, he may present traces of specific disease in other parts of the central nervous system. The chief difference, according to this, between the ordinary forms of myelitis and the specific diseases of the cord can be understood if we remember that the symptoms of an acute or subacute myelitis prove that the entire cross-section of the cord is affected almost simultaneously and to an equal degree, whence it follows that in such cases severe paralysis is likely to be associated with severe anesthesia, with marked contractures, with absolute loss of vesical and rectal control, with serious trophic disturbances, and so on; whereas, in the cases of spinal syphilis the morbid process invades the cross-section of the spinal cord partially and slowly. We may, therefore, find symptoms which point to a very marked affection of one or more of the systems of the spinal cord and to relative immunity from disease of the gray matter or other portions of the cord. Furthermore, in the ordinary cases of myelitis the symptoms point to a certain portion of the spinal cord at which the disease is most intense, while in cases of syphilis of the spinal cord the clinical symptoms show that the disease involves a very large portion, if not the entire spinal cord, yet affects each single segment relatively little. A still further aid to differential diagnosis is the very frequent involvement of the brain, at the same time that the majority of the symptoms point to disease of the spinal cord, or if these two sets of symptoms do not set in simultaneously, we frequently have in a patient who presents symptoms of a chronic specific myelitis the history of a preceding illness in which the symptoms were of a cerebral, rather than of a spinal, character. . . . The rapid and often unexpected recovery,

as well as the relapses, help also to distinguish these cases from the usual forms of myelitis."

**Treatment.**—All of the preceding forms of syphilitic disease (with the possible exception of Erb's type) are amenable to vigorous antisyphilitic treatment. Let us suppose that in a given case the blood or the cerebrospinal fluid or both have given the well-known indications of syphilis. Unless there are serious contra-indications, treatment by salvarsan should be instituted promptly. Among the contra-indications are: (1) Very advanced age (with its brittle arteries and tendency to hemorrhage); early infancy; (2) acute or chronic nephritis; (3) diabetes; (4) myocarditis, and advanced arteriosclerosis; (5) advanced optic atrophy (in progressing optic neuritis great caution should be exercised). We cannot agree with Osler and Churchman<sup>1</sup> that nervous lesions are a contra-indication. After apoplectic seizures and in basilar or cerebellar lesions, great caution should be exercised; in the late stages of general paresis and of tabes the treatment may be fruitless, but in all other nervous conditions of specific origin salvarsan treatment is amply justified.

Mention should be made of the neuro-recidives as a possible contra-indication for repeated salvarsan injections. Curiously enough, we have seen far less of it in this country than was reported abroad. It is fair to suppose that the salvarsan injections may have acted as a provocative agent, and these recurrent nerve lesions are the expression of a latent syphilis. The experienced physician or neurologist will not desist because of these nerve lesions, unless the symptoms point to the respiratory or cardiac centres, in which case great caution must be exercised.

**Plan of Treatment.**—Salvarsan should be administered by intravenous injection; by common consent the intramuscular injections have been abandoned. In all adults, especially in women, 0.3 gm. is the initial dose, to be repeated after a week or ten days. If the first injection has been well tolerated, 0.4 or 0.5 gm. salvarsan may be given at the second treatment. Allow an interval of four weeks to elapse, during which period one intramuscular injection of one grain of the salicylate of mercury may be given each week. Unless the symptoms are very urgent, it is well to give the patients a complete rest from all treatment for another fortnight, and then repeat the course of salvarsan and mercurial injections. At the end of the second course of injections, it will be well to examine the blood and spinal fluid again, or, at least, the former, to determine whether or not the treatment has had any effect upon the constitutional symptoms. Dreyfuss has shown that there is more danger in too little, than in an excess of, anti-syphilitic treatment.

In cases of subacute or chronic syphilis of the central nervous system (specific headaches, recurrent attacks of specific meningomyelitis, recurrent apoplectiform seizures) it is well to advise the patient to submit to repeated courses of treatment every six months.

The *technique* of salvarsan injections should be within the grasp of every physician. After several years of innumerable modifications, it has become relatively simple. A very large personal experience has impressed

<sup>1</sup> The article on Syphilis (in Vol. II) has many excellent hints on the subject.

upon the writer the importance of preparing the salvarsan solution with the utmost care.<sup>1</sup> *Freshly distilled water* is the first essential. Next in importance comes the accurate neutralization of the solution which should be introduced at a few degrees above body temperature. More important than all else is the careful introduction of the needle, the point of which must lie safely within the lumen of the vein. If the needle is correctly placed, the patient does not experience the slightest pain. It is well to go very slowly at first. If the injection is in the least painful, withdraw the needle at once and completely. Try another vein, or, if necessary, the other arm. The smallest amount of the solution poured into the perivascular tissue will give rise to disagreeable induration. With increasing experience, the use of normal solution before introducing the salvarsan has become superfluous; we have also learnt to use more concentrated solutions (0.3 gm. to 40 cc.). If the physician is certain of the position of the needle, the solution may be injected with an ordinary Luer syringe, without preceding or subsequent saline solution.

Neosalvarsan has been widely used. It is more easily dissolved; cold (sterile) water must be used; the dose is twice as large as that of salvarsan. Excepting the ease of preparation, it has no advantages over the older drug. Its equal efficiency is a matter of doubt; those who are familiar with the technique should use the original drug.

Salvarsan injections may now be given in a physician's office or operating room. The patient and the physician must have ample time, and the patient should be required to rest at least one hour after treatment. A light diet and plenty of rest and quiet for the first twenty-four hours should be urged.

In this era of the dominance of laboratory methods, it has been urged that salvarsan treatment be continued until the Wassermann reaction of the blood and of the spinal fluid becomes negative, and until the cell count of the spinal fluid is much diminished. The reduction of the cell count may be brought about, in many instances, often without the slightest change in the clinical manifestations; the Wassermann reaction, particularly of the blood, is often difficult to change; these are "Wassermann fast" reactions; and in such patients it is safer to be guided by the change in clinical conditions, than by a change, or absence of change, in biological conditions.

**Intraspinal Treatment.**—With the knowledge that the central nervous system is frequently the seat of an active luetic process, came the desire to introduce some spirocheticidal substance directly into the spinal canal. Swift and Ellis introduced the use of salvarsanized serum by injection into the spinal canal. They were able to bring about marked changes in the cerebrospinal content and reported considerable improvement, particularly in cases of tabes dorsalis, and some improvement in general paresis. The Swift-Ellis method consists in giving a patient an intravenous injection of 0.5 gm. of salvarsan. One hour later sufficient blood is withdrawn to yield about 15 cc. of serum. The next day this serum, carefully prepared, is injected at body temperature into the spinal canal.

<sup>1</sup> The details of the technique are well given in Kaplan's book, p. 213 *et seq.*

The quantity of spinal fluid removed is to equal the quantity of serum-salt mixture introduced.

For a short period the salvarsanized serum method met with considerable favor. If the patient's own blood-serum was surely "salvarsanized" it was natural to consider the Swift-Ellis method full of hope. It would make it possible to bring the arsenic preparation into very close contact with the specific tissues within the spinal canal. The discomfort occasioned by the intraspinous procedure is considerable; the method is also not entirely devoid of danger. It was necessary, therefore, to inquire whether this method accomplished much more than did the much simpler and less dangerous intravenous method. In association with Strauss and Kaliski,<sup>1</sup> the present writer gave considerable study to this problem. "It seemed . . . that blood withdrawn three-quarters of an hour after an intravenous injection would have to depend for whatever efficiency might be attributed to it upon the presence of the salvarsan itself, since in the short interval . . . the organism could not elaborate any form of protective bodies." As a matter of fact, Prof. Benedict—a chemist of unquestionable authority,—helped us to determine how little salvarsan was actually circulating in the blood. A man weighing about one hundred and fifty pounds and having a total blood volume of about eight pounds, if injected with 0.4 gm. of salvarsan would have in every cubic centimeter of his blood immediately after the injection, before elimination or fixation of the body cells began, about 0.0001 gm. of salvarsan. This was the maximum amount that Benedict found in 20 cc. of whole blood. It was shown that the arsenic is not free in the blood stream but is probably bound by the receptors of the body cells.

Benedict also showed from an analysis of four specimens of spinal fluid withdrawn twenty-four hours after an intravenous injection of salvarsan (0.4 gm.) that the spinal fluid contains free arsenic in about one-sixth to one-tenth the concentration in the whole blood. This very striking fact is opposed to the usual belief that none of the drug administered intravenously finds its way into the spinal canal. If the reduction of the fluid cells is the one desideratum, Mitchell<sup>2</sup> has shown that this can be accomplished by lumbar puncture, frequently repeated.

Marinesco<sup>3</sup> and Wechsleman developed a method (modified by Ravaut) of intraspinous administration of neosalvarsan. Ravaut uses a 6 per cent. solution of neosalvarsan in water. Each minim would contain approximately 0.003 gm. of neosalvarsan. The needle is introduced as for lumbar puncture; the spinal fluid allowed to run into the funnel of the apparatus, from one to four drops of the solution are added as desired, and the mixture then allowed to run by gravity into the spinal canal. Kaliski has modified this method so that each 0.1 cc. of a salt solution contains 0.001 gm. of neosalvarsan.

The proper administration of salvarsan will require further study, and the elaboration of newer methods. Meanwhile many cases will call for the occasional use of mercury and even the iodides. Very recently Joseph Byrnes published a method by which mercury was injected intra-

<sup>1</sup> *Am. Jour. Med. Sc.*, 1914, cxlviii, 693.

<sup>2</sup> *Jour. Nerv. and Ment. Dis.*, 1914.

<sup>3</sup> *Ztschr. f. diätet. und phys. Therapie*, 1913, xvii, 194.

spinously. His results were surely as satisfactory as those obtained by other intraspinal methods.

Mercury should be given in the form of inunctions, or intramuscular injections of the salicylate of mercury ( $\frac{1}{2}$  to 1 gr. per week), or in the form of the old-fashioned "mixed treatment." The iodides,—the mainstay of former decades—may be given in saturated solution, fifteen to fifty drops, well diluted, three times daily

### TABES DORSALIS.

**Synonyms.**—Tabes dorsalis is also known as locomotor ataxia, and posterior spinal sclerosis, "Rückenmarksschwindsucht" of the Germans.

**Definition.**—Tabes dorsalis is a chronic, more or less progressive disease of the central nervous system, exhibiting its chief morbid changes in the spinal ganglia, in the posterior roots and posterior columns of the spinal cord; it is characterized clinically by a very definite series of symptoms, among which the Argyll-Robertson pupil, the lightning pains, the girdle sensations, the loss of the deep reflexes, the hypotonia, and the ataxic gait are the most prominent.

**Etiology.**—During the last third of the nineteenth century the neurologists were divided into two camps with reference to the etiology of tabes dorsalis. The one faction, led by Fournier and Erb, claimed that tabes dorsalis was a true syphilitic affection of the spinal cord; the other faction, led by Leyden, believed that syphilis was only one of a number of different causes that might produce this disease. The latter accused the adherents of the opposing theory of attaching undue importance to syphilis. As a matter of fact, the statistical data gathered by Erb and his pupils proved that in about 95 per cent. of the cases of tabes dorsalis there had been preceding syphilitic infection, and we may add to this that when the disease afflicted women, as it does much more rarely than men, the percentage of preceding syphilitic infection is even higher. The writer has for years paid the closest attention to this point, and in his own private practice, he has rarely seen a case of tabes in which the etiological factor of syphilis could be excluded. In only one case was he certain of this, and that was in a patient who had been entirely well until the age of thirty-five, and had never exhibited the slightest sign of syphilis, nor had he contracted any infection. He was struck with a piece of shell after a dynamite explosion, and within six weeks of that time developed the first symptoms of what proved to be a rapidly progressive form of locomotor ataxia. More recently the Wassermann reaction and the study of the cerebrospinal fluid have revealed the tremendous importance of syphilis in the etiology, the percentages of positive blood findings varying from 90 per cent. (Nonne, Wassermann and Plaut) to 76.6 per cent. (Sachs, Castelli). The lower percentages recorded by the latter may, perhaps, be due to the established practice in this country of subjecting tabic patients to prolonged antisiphilitic treatment. Erb's dictum of years ago, that a person who has not had syphilis was not likely to have tabes dorsalis, has been fully substantiated by later

experience, and while trauma and possibly extreme exertion<sup>1</sup> may be occasional factors, we are safe in stating that syphilitic infection is the almost universal cause of tabes dorsalis.

Locomotor ataxia appears, as a rule, five to twenty years after the initial infection. If syphilis is acquired late in life, syphilitic forms of spinal disease may appear within a very few years. The writer has seen the first symptoms of tabes dorsalis, including optic atrophy, in a man, aged fifty-five years, appear within one year of the initial infection.

**Pathology.**—The chief anatomical change is a degeneration of the posterior columns of the cord. Even the gross specimen reveals a distinct grayish discoloration of these parts. Under the microscope it is seen that the degeneration begins in the columns of Burdach at the level of the upper lumbar segment. It must be noted that this is the area through which the posterior roots pass. At higher levels the columns of Goll are also involved at a very early period of the disease. At a later stage the entire posterior columns of the lumbar and dorsal segments are completely degenerated. In the cervical segments the degeneration may be limited to the columns of Goll; later on here, as elsewhere, the greater part of the posterior columns is degenerated. In the morbid process the nerve trunks are degenerated, while the glia may remain either normal or may proliferate. The disease is not strictly limited to the posterior columns. The columns of Clarke and the posterior horns (particularly Lissauer's root zone) are frequently involved. The posterior roots are diseased so often that many investigators (Leyden, Redlich, Oppenheim) believe the changes in the spinal cord to be due to early involvement of the posterior roots. Oppenheim,<sup>2</sup> and Thomas and Hauser<sup>3</sup> have traced this degeneration to the spinal ganglia, the trophic centres of the posterior roots. The morbid process may extend into the medulla oblongata, involving the spinal root of the trigeminal nerve (thus accounting for the rare and interesting forms in which the trigeminal symptoms appear early in the disease). The solitary bundle, the sensory motor nuclei of the fifth nerve, the auditory nucleus (rarely), the hypoglossal nucleus, and the nucleus of the vagus nerve may become involved. In addition to all of these and much more frequently, we may find a degeneration of the optic nerve (simple white atrophy). Other cranial nerves may undergo simple atrophy. Oppenheim found a degeneration of the vagus and recurrent laryngeal and the glossopharyngeal nerves. Obersteiner and Schiff accounted for the hemiatrophy of the tongue by a degeneration of the hypoglossal. The auditory nerve is, of all cranial nerves, the one most rarely affected. Oppenheim's finding of a degeneration of the Gasserian ganglion is related to the involvement of the sensory root of the fifth nerve. Atrophy of the sensory nerves of the skin has been shown to occur.

All the changes enumerated refer to the sensory fibres of the periphery of the spinal cord. French writers (Charcot, Pierret, Raymond, and others) have shown that in some cases the anterior gray horns are also

<sup>1</sup> Poisoning by lead and ergot produces clinical and anatomical conditions similar to tabes dorsalis.

<sup>2</sup> *Archiv. f. Psychiatric*, vol. xviii, vol. vii, 44.

<sup>3</sup> *Nouv. Icon., etc.*, 1904.



involved. This will account for some of the cases of tabes with muscular atrophy, although it is doubtful whether the peripheral nerves may not be responsible for some of these muscular wastings. In conclusion, we may state that while the morbid process is essentially a posterior spinal sclerosis, it may involve other parts of the cord and of the central nervous system, as well as the peripheral nerves. The toxic agent may exert its effects upon almost any or every part of the nervous system, although it has certain definite sites of predilection; but to make tabes dorsalis a cortical disease because cortical fibres undergo atrophy (according to Jendrassik) is carrying the argument a little too far. All these changes may be secondary to changes in the central sensory neurones (Strümpell). The opponents of the Fournier-Erb doctrine of the specific origin of tabes dorsalis have urged that there is nothing characteristically syphilitic about the morbid process; there is some truth in this, but the force of this objection is lessened by the fact that tabes dorsalis does occur in association with a typical specific meningitis.<sup>1</sup> McIntosh and Fildes in their recent article (1914) express their surprise that so little has been made of this specific meningitis. It is generally accepted that Redlich and Obersteiner were correct in stating that the degeneration of the columns begins at the "ring of Obersteiner"—an apparent constriction in the posterior root at its point of insertion into the cord. Some have suspected early involvement of the cells in the posterior root ganglia, but this has not been proved. Nor is it unlikely that the specific leptomeningitis, found in some cases, is the invariable starting point of the tabic process. The early involvement of the meninges and of the roots would account for the earliest clinical symptoms. Noguchi<sup>2</sup> has proved the presence of the *Spirochaeta pallida* in the posterior columns.

**Symptoms.**—With the possible exception of general paresis, there is no disease presenting such a multiplicity of symptoms, and it is doubtful whether any one patient ever presents even a bare majority of the symptoms. The clinical picture will be described as it is outlined in the writer's mind from a large individual experience, even if there be some departure from the common description. First of all, there are two distinct groups, one the ataxic and the other the ophthalmic. The ataxic group represents the classical type. In this group of cases the optic nerve symptoms are developed very late or not at all, while all the ataxic and sensory symptoms attain to fullest development. In the ophthalmic group optic nerve atrophy is one of the earliest and most pronounced symptoms of the disease, leading to early blindness, while the ataxic symptoms in this group are very imperfectly exhibited. While the latter group presents fewer symptoms, the early development of amaurosis makes it the form most to be dreaded. Both groups have, however, a majority of symptoms in common. It has been customary to divide the symptoms into (1) those of the prodromal or pre-ataxic stage, and (2) those of the ataxic stage. The prodromal stage may last for months or years, and yet the final diagnosis cannot be safely established until

<sup>1</sup> See Nonne's monograph and the writer's article in the *New York Med. Jour.*, 1894, lix.

<sup>2</sup> *Münch. med. Wchnschr.*, 1913, 737.

several of the cardinal symptoms have appeared. The presence of three of the cardinal symptoms is sufficient for this purpose, and among those cardinal symptoms we must rank subjective as well as objective signs.

The cardinal symptoms of *tabes dorsalis* (given approximately in their order of importance) are: (1) Lancinating pains, (2) Argyll-Robertson pupil, (3) loss of deep reflexes, particularly of the knee-jerks and of the Achilles tendon reflex, (4) Romberg symptom (swaying of the body with the eyes closed). (5) Girdle sensation in various forms; hyperesthesia as well as delayed sensation. (6) Hypotonia of the muscles. (7) Bladder disturbances (rectal insufficiency is rare). (8) Ataxic movements of the lower extremities. (Ataxia of the upper extremities is relatively rare.) (9) Sexual weakness. (10) Cranial (more particularly ocular) nerve palsies, strabismus, double vision, etc. (11) Optic nerve atrophy, single white or primary white atrophy. (12) Visceral crises. (13) Trophic disorders.

Any combination of these symptoms is possible, but the majority of cases are characterized for a long period of time by lancinating pains, some forms of hypesthesia, pupillary symptoms, and by the loss of the deep reflexes. After the lapse of some time the ataxia and the bladder symptoms appear.

The *lancinating pains* occur, according to Erb, in 90 per cent. of the cases, and in 70 per cent. of all cases of *tabes* they constitute the earliest symptom. They are lightning-like, severe, darting, or boring pains occurring in any part of the body, most frequently down one or both legs within the distribution of the sciatic, around the calves, and into the heel or toes. Sometimes they are particularly annoying within the distribution of the crural nerves. These lancinating pains occur less frequently in the upper extremities, are localized often enough in the thorax and sometimes even in the face (*tabes superior*). The lancinating pains are so commonly sciatic in distribution that persistent sciatica, particularly if bilateral, is suggestive of *tabes*. These lancinating pains come on without reason when the patient least expects them. Any slight change of temperature may be sufficient to produce them.

*Hyperesthesia* may be noted within the special areas of pain, so that the pressure of the clothing or of the bedclothes becomes intolerable. Closely allied to the subjective sensory disturbances just recorded are various forms of paresthesia, such as formication, a feeling of "pins and needles," a feeling as if the patient were walking on velvet or cotton, peculiar feelings of tension about the perineum and the genitals. Formication in the distribution of the ulnar nerve is the commonest among those in the upper extremities. Disturbances of sensation around the thorax give rise to the well-known girdle sensation—a tolerably early and very characteristic symptom. These sensory disturbances are often the initial symptoms, and can be accounted for by the early involvement of the posterior root fibres and their prolongations into the cord. Tabic pains are, to a degree, early root symptoms.

The *Argyll-Robertson pupil* occurs in at least 80 to 90 per cent. of all cases. It implies a failure of reaction to light, whereas the pupils react promptly during convergence and accommodation. Simple as the

phenomenon is, experience is necessary to determine the exact manner of pupillary response. It is well to examine the patient in a darkened room with artificial light (many dark pupils cannot be examined by ordinary daylight). Unless every attempt at convergence is excluded, a false inference may be drawn. Also remember that the phenomenon applies only to eyes with normal or nearly normal vision.

The *immobility of the pupil* to light is generally bilateral. Differences in the reaction of the two pupils may be observed in the earliest stages. At other times it is strongly suspicious of an active luetic process. Tabic pupils are generally miotic ("pin-hole pupils"). Even the smallest pupil may contract still further during convergence, but will not do so under the stimulation of light. The contours of the pupil are apt to be irregular. Oculists lay less stress upon this, but neurologists are generally agreed with the writer in holding this sign to be one of importance in tabes, and particularly in those cases in which the luetic virus is still active. Mydriasis occurs in those cases in which there is complete immobility of the pupil, and may be unilateral. In the writer's experience the Argyll-Robertson pupil is the earliest objective, and by far the most constant, symptom. A close second is the loss of the deep reflexes.

The *loss of the patellar tendon reflex* (Westphal's phenomenon) is one of the earliest and most constant symptoms. It is present in fully 95 per cent. of the cases, although in a few instances, particularly in the earlier stages, the knee-jerk may be either present in one or both legs, or may still be elicited by reinforcement (Jendrassik's method). It is not unusual to observe a gradual disappearance of the knee-jerks with the development of other symptoms. For this examination the patient should be tested when the knees are bare, else mistakes are bound to occur; and if there is any doubt it is wise to put the patient in a recumbent position with the knees semiflexed, or else to lay the patient flat on a couch or bed with the legs hanging down over the edge. At times a diminished knee-jerk can only be discovered by placing one hand on the quadriceps while tapping the tendon with the other. A hypotonic condition of the quadriceps is a more or less constant accompaniment of the loss of the knee-jerk. It is a well-known fact that many persons who know the value of this symptom inhibit the reflex more or less unconsciously.

Of late years we have come to regard the *loss of the Achilles tendon reflex* as being of equal importance with the loss of the knee-jerk. Some writers even claim that it often precedes the loss of the patellar tendon reflex. This is surely not the rule. The loss of the Achilles reflex is of especial value if it is present on one side and absent on the other.

Loss of the deep reflexes of the upper extremities is not as valuable a symptom, for the simple reason that these reflexes are not invariably present in normal individuals. The triceps tendon reflex is the most constant, but failure to elicit it cannot be considered a diagnostic aid unless other deep reflexes are also wanting, except possibly in the few (and rare) cases of cervical tabes. The unilateral loss of the triceps or wrist reflex would be significant.

By contrast the *superficial reflexes* remain unaltered, disappearing in

those cases only in which there are very grave disturbances of cutaneous sensation. The plantar, the cremasteric, the gluteal, and the abdominal reflexes can be elicited in tabic patients easily enough. The Babinski phenomenon is never present, as it is absent also in all normal persons.

The next most important and very constant sign is the *Romberg symptom*—the swaying of the body with the eyes closed. This is due chiefly to the loss or alteration of superficial and deep sensibility. To elicit this symptom the patient is asked to stand with eyes closed and with the feet touching one another at the toes. Erb has shown that this necessitates careful balancing of the body, and can be carried out only if the patient appreciates exactly his position in space and his contact with the floor. The sensation of the skin of the feet must be normal and the patient must also have full knowledge of the tension of his muscles and ligaments, or else he cannot maintain his proper static equilibrium. So long as the patient is able to control by vision his whereabouts in space, he may be able to stand perfectly; with the removal of visual control the sensory defects become evident. This test should be repeated a number of times; even normal individuals on a first examination sway considerably when asked to close their eyes. It is rare to find the Romberg symptom in tabic patients with optic nerve atrophy and blindness. Some years ago the writer reported the case of a blind tabic individual who swayed when he closed his blind eyes. This was evidently a psychic reminiscence of his early tabic period.

In connection with the Romberg symptom brief reference may be made to the *sensory disturbances*. In the vast majority of tabic patients there is, at least in the earlier stages, a very slight change of tactile sensation. At all events, nothing more than a moderate tactile hypesthesia exists. Such changes as do occur are most common in the soles of the feet, on the inner or outer margin of the feet, and within the distribution of the ulnar nerve. Starr<sup>1</sup> has given excellent illustrations, taken from Bonar, of areas of anesthesia in cases of locomotor ataxia. They bear all the earmarks of true segmental anesthesia, but few cases present this symptom in such marked form.

The *pain sense* is more often affected. Hypalgesia and delayed pain sensation are the rule. The latter is a very striking phenomenon, the patient perceiving it as only a mere touch (although pricked with a pin), and after a second or more perceiving (at times) intense pain. Erb has shown that faradic sensibility is much diminished. On the whole, this is proportionate to the loss or diminution of the general pain sense.

The *temperature sense* is apt to be altered. The perception of heat may be diminished and cold felt more acutely than by normal individuals. Exceptions to this rule are not infrequent. Allochiria, reference of a sensory stimulus to the opposite side of the body, the writer has observed a number of times; polyesthesia, the perception of several sensory impacts when touched but once, has also been recorded, but the writer has not seen it except as a species of self-deception. Astereognosis occurs in the

<sup>1</sup> *Organic and Functional Nervous Diseases*, third edition, 1909, p. 352 *et seq.*

advanced stages, also analgesia of the genitals in both sexes; both are, however, rare phenomena.

Sensory disturbances of the trunk of the body are present in a very large number of tabic patients. These hypesthetic and anesthetic zones are developed early, giving rise to various forms of girdle sensation, showing a distinct root and segmental distribution. These forms of disturbed sensation are symmetrically developed, and if they are asymmetrical they correspond to the anatomical distribution of the earliest tabic (root) changes in the dorsal segments. If the lower part of the spinal axis is involved, we are apt to find similar paresthesia and hypesthesia around the perineum and genitals.

Disturbances of *deep muscular sensibility* are largely responsible for the Romberg symptom. They may be revealed by other tests. The patient has lost all power to appreciate the exact position of his limbs. If he is in the recumbent position and is asked to elevate his leg to the exact level at which the examiner has placed the other leg (the patient's eyes being closed), he is unable to do this. He may also fail to appreciate simple passive movements of the joints. All these tests can be carried out more readily with the lower than with the upper extremities.

Tabic patients are supposed to have an impaired sense of fatigue (a very inconstant symptom). It is certain that they have a diminished osseous sense, a diminution or loss of vibratory sensation.

A most valuable, reliable, and early symptom of tabes is a distinct *muscular hypotonia*. Erb, Leyden, and Frenkel have studied the phenomenon carefully (Frenkel introduced the term). It may and often does precede the ataxia, and is, therefore, of great diagnostic importance. Not infrequently it helps to establish the diagnosis. It consists in an abnormal diminution of the tension of the muscles and ligaments, so that excessive passive movements of the joints are possible. The normal individual can flex the thigh upon the trunk with the leg extended 50 or 60 degrees; the hypotonia of tabes permits this flexion to 90 degrees or more. Hyperextension of the knee in the recumbent position and genu-recurvatum in the erect position are constant phenomena. The same freedom of joint movement (according to Frenkel) is to be observed in the spinal column also in the joints of the hands and fingers. The abdominal and spinal muscles also are in a condition of hypotonia. The condition is independent of the degree of ataxia and of the loss of deep sensibility. It has a considerable influence on locomotion.

We pass now to the consideration of the *ataxia* which is so prominent a symptom of the disease. We have hinted above that there are some cases not ataxic from the beginning to the end of the disease. Ataxia implies not loss of gross muscular power, but an inability properly to coördinate muscular movements. In more than nine-tenths of the cases ataxia appears first in the lower extremities, and very insidiously at that. The patient notices at first that he has become a little unsteady on the legs, that he walks as though he were intoxicated, that his legs "give way" under him. He experiences great difficulty in going up or down stairs; he stumbles in jumping off or on a car; cannot cross the street in safety; has to watch his movements; must look at his feet or else he does not

know where they are; if he is to walk at all, he must walk with a broad base and can walk with a stick only; finds much more difficulty in walking at night than during the day, and stumbles or falls if he tries to get out of bed in the dark. By degrees he has developed the typical tabic gait.

These earlier stages of tabic ataxia are much more manifest on examination. If the patient is asked to stand on one foot, he topples or falls; if asked to turn about on his feet quickly, he sways badly; if asked to walk a straight line, placing one foot in front of the other, he is utterly incapable of doing it; in all movements requiring coördination of the muscles he goes wide of the mark. In the recumbent posture he cannot elevate the leg when extended without performing zigzag movements; he has difficulty in touching one knee with the opposite heel, and with the eyes closed all these defects are much exaggerated, particularly if there is considerable impairment of deep muscular sensibility.

Many patients learn to accommodate themselves to a certain degree of ataxia, and for years (particularly under the influence of modern therapeutic methods) get about with considerable ease. As the disease progresses the increasing ataxia renders the limbs useless and the patient becomes chair or bedridden. In extreme cases the patient, even if supported, has lost control over his legs so completely that, on attempting to walk, the legs are thrown about in the wildest, often grotesque, fashion. Ataxic movements of the upper extremities are, on the whole, very rare, and there are few who have lost the ability to write or feed themselves. In that rare form known as *tabes superior* the ataxic movements of the hands are more pronounced. The ordinary movements of daily routine become impossible. Ataxic movements of the facial muscles, of the lips, of the tongue, of the laryngeal muscles, are rare.

These are the most typical and constant symptoms, but there are a number of others, the student remembering that their absence need not militate against the diagnosis of tabes.

*Vesical Disturbances.*—These are important and often most troublesome. They are so common, even in the earliest period, that they not infrequently constitute the first symptoms that arouse a suspicion of serious organic nervous disease. In the genito-urinary service of a large hospital the neurologist is often consulted for the purpose of determining whether or not vesical symptoms, which appear to be independent of disease of the bladder itself, are the expression of a latent (?) tabes. At first dysuria (neuralgic pain in the region of the bladder), hyperesthesia of the neck of the bladder, dribbling, frequent micturition or moderate retention, are the usual forms of disturbance. As the disease progresses the amount of residual urine increases; catheterization may be necessary and cystitis is only too apt to be superadded. This increases the discomfort, and if it leads to pyelonephritis there is distinct danger to life.

*Rectal insufficiency* is rare. In some patients with hypesthesia of the anal region inconvenience is experienced after taking purgatives, but otherwise the rectal sphincter continues to perform its function satisfactorily. The patient is more depressed over the loss of sexual desire and of sexual power, the latter often preceding the former. Female

tabic patients are reported to experience a loss of sexual desire, but the point is not easily established.

*Cranial nerve* disease is an important factor. Oculomotor palsies, partial or complete, abducens paralysis, rarely trochlearis palsy, each and all are known to occur. The palsies of the earlier period—ptosis and strabismus—are particularly significant, because they occur even more frequently as the symptoms of cerebral lues. The diagnosis between tabes dorsalis and cerebral syphilis is established by the association of other characteristic symptoms of locomotor ataxia. In the more advanced stages we may find paralysis of associated movements of one or both eyes, pointing to involvement of the various nuclei. Complete ophthalmoplegia externa and interna are observed in rare instances.

The *facial nerve* is so rarely involved that mention of this fact would not be necessary if facial nerve symptoms and those due to involvement of the motor branch of the fifth nerve did not occasionally occur in rare forms of tabes complicated by bulbar disease. The sensory branch of the fifth nerve is not as exempt as is the motor division of this nerve. It has been my peculiar experience to see a number of patients in whom a dissociation of sensation, at first unilateral and later on bilateral, within the distribution of the trigeminal nerve, has been the earliest symptom of tabes. In two of these patients this fifth nerve dissociation preceded all other symptoms by many years. During this period the diagnosis was much in doubt. The suspicion of an unusual form of syringomyelia was entertained until the cardinal symptoms of tabes left no doubt.

Palsies due to involvement of the *tenth* and *eleventh* nerves also occur, but rarely, if at all. The most frequent form among these is characterized by dyspnoea and inspiratory stridor. One of Erb's patients had to have tracheotomy done, and wore a cannula for six years. Hoarseness and aphonia, due to paralysis of the recurrent laryngeal nerve, have been recorded. Atrophy or paralysis of the trapezius and sternocleidomastoid muscles, due to involvement of the eleventh nerve, is excessively rare. A little more frequent is the atrophy of the tongue due to disease of the hypoglossal nerve, but this condition is associated, as a rule, with other symptoms of bulbar paralysis. Progressive cranial nerve palsies, if not a typical progressive bulbar paralysis, may occur in association with the other symptoms of tabes. The involvement of one or more, if not of all of the nuclei of the cranial motor nerves, may cause an endless variety of symptoms. The olfactory and auditory nerves escape, as a rule, although cases have been described with loss of the sense of smell, with delusions of smell, and still others in which there has been continuous tinnitus with vertigo. Ménière's symptom group has also been recorded.

But by far the most important cranial nerve symptom is the *optic nerve atrophy*. This has been referred to above, where it was said that it gives rise to a distinct group of cases known as amaurotic tabes.

Among the orthodox symptoms, although often wanting, are the *visceral crises* and the *trophic disorders*. We owe our knowledge of the visceral crises largely to the accounts given by Charcot. In his inimitable way he described gastric crises, and nowadays every neurologist recognizes the condition. But they still seem to be a *terra incognita* for the general

practitioner and for the specialist in gastric and intestinal disorders. The gastric crises consist of severe neuralgic pains in the epigastrium or in some other part of the abdomen, appearing suddenly, lasting for hours or days and then disappearing quite as suddenly. The intense pain is accompanied by uncontrollable vomiting. It will be readily understood why this condition leads to errors in diagnosis.

Entirely similar to the gastric are the *intestinal crises*. One patient with trigeminal dissociation also suffered from frequent inexplicable diarrhœa with intense abdominal pain, until the proper interpretation of both was made manifest by the appearance of a series of tabic symptoms.

Among the curiosities of tabetic symptomatology we may class the vesical, the renal, the testicular, and even the clitoris crises. Special mention must be made of the laryngeal crises. These begin with a tingling sensation in the throat, a feeling of constriction leading to a condition of temporary asphyxia with stridor, cyanosis, and occasionally to a convulsive seizure. The spasm lasts but a few minutes, when the patient is out of imminent danger. Oppenheim has described pharyngeal crises with pain in the pharynx and frequent movements of swallowing.

*Cardiac crises* are referred to because the writer has seen them in several cases, characterized by intense pains, like those of angina pectoris; but as these patients were at an age when angina pectoris could not be excluded, their exact value as a symptom of tabes is a matter of dispute.

Lastly, a few words about *trophic disorders*. The best known among them is the perforating ulcer, which is generally situated below the big toe at the level of the metatarsophalangeal joint, but it may occur in other parts of the sole of the foot. It is a small, circular ulcer, extending through the soft parts to the bone. It does not yield readily to treatment. Erb claims that it is cured more often by mercurial salve than by anything else, and for this reason he believes it to be specific in character. Herpes zoster has been observed often enough to make it worthy of special mention.

Charcot and his pupils were the first to dilate upon the *arthropathies*. The knee-joint is the one most frequently affected; next in frequency come the hip- and shoulder-joint, but marked and similar trophic changes occur in the elbow- and finger-joints. The chief characteristic is an enormous swelling of the joint with little or no increase of synovial fluid. The joint is, however, much enlarged, remains normal in color, and absolutely painless. The arthropathies may appear early, and if they become chronic lead to grotesque deformities, to luxations and subluxations, to the formation of osteophytes; the bones become unusually porous and the slightest accident may cause fracture in and around the joint. Painless fracture of the long bones on slight exertion or slight injury is tolerably frequent. In the ankle-joints and in the smaller joints of the foot similar structural changes occur that Charcot described as the tabic foot—a grossly misshapen foot. A limb or joint so affected becomes thoroughly useless, and yet it is astonishing how long patients with these arthropathies manage to get about. Not unlike these arthropathies are the alterations of the alveolar processes leading to a loss of



teeth. All the teeth may drop out in this way. It is so rare that in hundreds of cases of tabes the writer has not seen a single instance.

Much has been made of the occurrence of *cardiac disease* in tabes. When it does occur it is an accidental complication, and it is due to the preceding syphilitic infection. In some investigations of cardiac conditions, by Collins and the writer,<sup>1</sup> with the Wassermann method, the specific basis was well established, as was done before by Citron and others.

Lastly, we must take note of the *psychic* manifestations. Conditions of depression, of neurasthenia, of morbid irritability, are not unusual or difficult to account for in view of the distressing condition. Sufferers from locomotor ataxia may develop paranoia or paranoid states. Some of them may pass through states of acute maniacal excitement or through states of transitory dementia with and without aphasia, probably an expression of brain syphilis. But all these psychic manifestations are relatively less frequent in tabes than the occurrence of symptoms characteristic of general paresis. Both diseases have a common etiology—syphilis. It is not astonishing, therefore, that some patients should develop both conditions. As a matter of fact, tabic patients do develop often enough the symptoms of general paresis (delusions of grandeur, loss of memory, parietic speech, etc.). Still more frequently do patients with general paresis develop a series of tabic symptoms (lightning pains, ataxia, loss of knee-jerks, etc.). Some symptoms, such as the pupillary reflex, immobility, loss of vesical control and of sexual power, both diseases have in common. In view of this community of symptoms, we need not hesitate to declare that tabes and general paresis are due to one and the same process, that both are metasyphilitic diseases, and that it is only the difference in localization that determines whether the morbid process gives rise to the clinical picture of tabes or of general paresis. That there is an intimate etiological relation between the two diseases cannot be denied, yet their simultaneous occurrence is, after all, relatively rare. Tabes followed by general paresis is distinctly rarer than general paresis followed by tabes. The association is rare enough to caution the physician not to make the diagnosis of general paresis whenever a tabic patient presents slight psychic abnormalities.

Before concluding, it may be well to recall the fact that the disease may last fifteen to twenty or even thirty years, unless life is terminated by some intercurrent disease. Of all the symptoms, blindness, lancinating pains, cystitis and the visceral crises are the most distressing. If the disease drags on, the ataxic limbs may become paralyzed; in this helpless condition the occurrence of bedsores, the incontinence of urine and feces, the increasing marasmus, make a truly pitiable picture. Fortunately patients are usually relieved of their misery by some intercurrent disease before these extreme conditions appear.

Bearing in mind the great multiplicity of symptoms, one can readily see that there may be a vast difference in the grouping of symptoms of various cases of tabes, and also of the general course in one or the other individual. In some instances the course is unusually slow, so that the

<sup>1</sup> *Am. Jour. Med. Sc.*, 1909, cxxxviii, 344.

disease appears to be stationary for ten to fifteen or twenty years; in others the disease may come on insidiously and suddenly take a turn for the worse, developing within the course of a few months a number of the most distressing symptoms. As a rule, the prodromal stage covers a period of months or years. By slow stages a slight difficulty in walking progresses into a marked ataxia. Only now and then the ataxia is developed with startling rapidity.

It has been customary to designate various types according to the predominance of some one set of symptoms or according to the localization of the disease as indicated by the chief symptoms. The terms explain themselves; thus, various writers choose to speak of *tabes dolorosa*, of *tabes paresthetica*, of *tabes atactica*, of *tabes paralytica*, and *tabes amaurotica*; other designations are *tabes visceralis*, and *tabes superior* (meaning cases in which the upper extremities are chiefly involved). *Tabes juvenilis* applies to those rare forms in which all the symptoms appear early in life.

Erb has made special studies of the cases to which he applies the term *tabes incompleta*. These are practically identical with the *formes frustes* of the French writers. In this special group only a few of the cardinal and characteristic symptoms are present. While this may be the rule in the earlier stages of many cases, the term should be applied only to those in which this paucity of symptoms has been noted for a number of years. There are individuals, who, for years, have had the Argyll-Robertson pupil, others who have had visceral crises, still others who have had slight vesical disturbances, and every one of them suggestive of tabes, and yet in the absence of corroborative symptoms the physician may well hesitate to make a final diagnosis of tabes.

**Diagnosis.**—It is evident that the fully developed forms can hardly be mistaken for any other affection of the nervous system. If two or three of the cardinal symptoms are present the diagnosis can be made without any hesitation, but it is well also to bear in mind that the disease may begin with one or the other of the more unusual symptoms, with visceral crises, with an arthropathy, with vesical and rectal disturbance. These rather unusual symptoms may precede the cardinal symptoms for months or years, and in such cases the exact nature of the disorder cannot be diagnosed until one or more of the cardinal symptoms appear. The early recognition is so important that the physician may be pardoned in venturing the diagnosis of tabes on mere suspicion and of subjecting the patient to such treatment as may possibly help to inhibit the progress of the disease. In the recognition of the earlier stages, evidence furnished by the Wassermann reaction and by examination of the cerebrospinal fluid will be of the greatest importance.

In spite of all precautions, mistakes will happen, and the question will often arise whether the condition is one of *tabes dorsalis* or one of those now to be mentioned. The other chronic systemic affections of the spinal cord, such as *spastic spinal paralysis*, *amyotrophic lateral sclerosis*, and the *spinal amyotrophies*, need hardly be considered. In the latter the pupillary phenomenon are wanting, the deep reflexes may not be absent, the knee reflex may be increased, all sensory disturbances are wanting,

and the entire clinical picture is wholly different from that of tabes. Some of the symptoms of tabes dorsalis may occur now and then in *tumors* of the cord, invading first the posterior half of the cord, and in *multiple sclerosis*, if the sclerotic areas happen to involve the posterior columns and the posterior gray matter rather than the pyramidal tracts, in which they generally occur. But in such cases in which there is a doubt, the occurrence of nystagmus, of altered speech, and the ataxic tremor will help to indicate the disease. Much more difficulty will be encountered in differentiating between a *syphilitic meningomyelitis* invading the posterior half of the cord and genuine tabes dorsalis. Many years ago the writer dilated upon the subject, to show that some cases which were considered to be genuine instances of tabes dorsalis were actually cases of specific infiltration of the meninges, starting in the meninges but invading the cord. The presence of such symptoms which point strongly to the true syphilitic nature of the process, such as complete immobility of the pupils, paralysis associated with the ataxia at an early stage of the disease, marked remissions and exacerbations of the symptoms, would help to indicate an active luetic process rather than a typical posterior spinal sclerosis. *Syringomyelia* would hardly come into question. In rare instances this disease, beginning in and affecting chiefly the lumbar portion of the cord, may give rise to symptoms suggestive of tabes dorsalis, but the dissociation of sensation and the absence of typical tabic pupillary symptoms will help to establish the differential diagnosis.

Much more difficulty will be experienced in differentiating between tabes dorsalis and the *combined sclerosis* of the spinal cord. In this condition we are apt to have an ataxic paraplegia, and at one or the other stage of the disease the ataxia may preponderate to such an extent that the patient may present so few of the spastic and paralytic symptoms that there will be little reason to suspect anything else than tabes dorsalis. If the ataxic symptoms are associated with increase of reflexes, and if the pupillary phenomena are not typical of tabes dorsalis, the diagnosis of ataxic paraplegia or of combined sclerosis of the cord may safely be made. *Friedreich's ataxia* is recognized by its occurrence early in life, by its family or hereditary disposition, by the absence of the characteristic pains and sensory disturbances of tabes, by the absence of vesical and rectal disturbances, and by the existence of normal pupillary reflexes. The ataxia of Friedreich's disease is also coarser and of a more awkward type, and it involves the upper extremities much more frequently than is the case with true tabes. The "heredo-ataxie cerebelleuse" of Marie is recognized still more readily by the exaggeration instead of the absence of the deep reflexes; by the presence of club foot, and a number of other signs which do not occur in genuine tabes.

From *cerebellar tumors* tabes dorsalis will be differentiated quite easily, for in the former the form of the ataxia, the vertigo, vomiting, double optic neuritis and the absence of pupillary immobility, of the Romberg symptom, and of the lancinating pains, and the much more rapid development of all the symptoms, will lead to the correct diagnosis. The greatest difficulty of all will be experienced on attempting to differentiate between tabes dorsalis and *alcoholic multiple neuritis*, or, as it was formerly called,

alcoholic pseudo-tabes. The resemblance to tabes lies in the ataxic movements of the lower extremities and in the absence of the deep reflexes, also in the occurrence every now and then of marked sensory disturbances. On closer examination the difference is much more marked, for in alcoholic pseudo-tabes the pupillary phenomena are not so constant, although there is often a sluggish reaction to light. There is almost invariably a considerable degree of paralysis with the ataxia, often also some muscular wasting. Lancinating pains and girdle sensation are not the rule in multiple neuritis due to alcoholism, and the entire development of the symptoms is more rapid than is the rule in tabes dorsalis. Alcoholic pseudo-tabes leads more commonly to a paralytic condition of the lower extremities, more often also than is the case in tabes to an involvement of the upper extremities, while the physical symptoms are less pronounced, and the inquiry into the habits of the patient will give assistance. In not a few instances the writer has been convinced that a moderate alcoholic excess was sufficient to produce a pseudo-tabes alcoholica which would not have been developed if the patient had not previously been infected with syphilis.

**Prognosis.**—This is either favorable or unfavorable, not so much according to the gravity of the symptoms, as it is proportionate to the general optimism or pessimism of the physician. It is true that it is practically an incurable disease. The pupillary phenomena and the loss of the deep reflexes are rarely if ever recovered from, and in this sense, after the symptoms of tabes have appeared, the patient is afflicted with the disease for all time. On the other hand, the disease is often very slowly progressive; it may at times be said to come to a standstill, and many a patient is able to attend to his routine duties and to enjoy a tolerably comfortable life for many years. There is much to be done for the tabic patient, and the latter may be given the assurance that there are many ways of making him fairly comfortable. The newer methods of treatment enable us to stay the progress of the symptoms for a considerable period of time; and the patients can be taught to use their ataxic limbs far better than was possible in former years.

Some years ago some French neurologists claimed that tabes dorsalis seemed to have taken on a milder form; they were inclined to attribute this to the more thorough specific treatment which the patients received. There is some doubt as to the truth of this, but the fact remains that we do not see as many of the distressing forms of extreme ataxia as in former years, and tabic amaurosis does not seem to be as common as it was some twenty-odd years ago. On the whole, cases of tabes in which lancinating pains are the earliest and severest of the symptoms, are not the most unfavorable. While these lancinating pains predominate, the other symptoms are slow to appear. The cases in which a marked ataxia appears at a very early period represent a group of cases which are apt to advance rapidly and to lead to a large number of complications, but even these can be favorably influenced by various forms of treatment. The writer has been particularly struck by the fact that the amaurotic type of tabes, while it is perhaps the most unfortunate one, is often more slowly progressive and many of these patients would have been tolerably

comfortable for fifteen or twenty years if it had not been for the loss of vision. The prognosis is most unfavorable in those cases with a cystitis or a pyelonephritis, in which other syphilitic phenomena, such as convulsions, have occurred as an early complication. The combination of tabes and general paresis is bound to lead, sooner or later, to a fatal termination.

**Treatment.**—The question of *prophylaxis* looms up prominently. By the time the diagnosis is safely established the spinal cord changes are so considerable that one cannot expect to do much more than to arrest the disease. No one has yet been able to claim that he can in any way influence by treatment sclerotic tissue in the spinal cord after it has been once fully developed. As regards prophylactic measures, all that can be expected is possibly to lessen the syphilitic contagion. Such a Utopian state of affairs is not within our reach, but we can at least demand of everyone who has contracted syphilis that he subject himself to prompt and sufficient treatment. While it would be difficult to prove that prompt and proper antisymphilitic treatment following upon the initial lesion prevents the development of syphilitic and parasymphilitic diseases of the central nervous system, it is at least probable that these serious manifestations could be to a degree lessened if energetic measures were resorted to in every instance at the outset of the disease. There is at least no doubt that when the first signs of any syphilitic or parasymphilitic affection of the central nervous system appear, energetic measures should be used. It is safe to establish the general principle that in every case of tabes dorsalis in which thorough antisymphilitic treatment has not been administered, such treatment should be given before anything else is attempted. At the present date, and in view of recent experiences, this statement may be modified to mean that if a positive Wassermann reaction has been obtained, the patient should receive prompt antisymphilitic treatment. If the reaction is negative, the same treatment may be given if there are clinical signs pointing to an active luetic process.

By appropriate antisymphilitic treatment the writer means salvarsan injections, mercury salicylate (gr. j by injection) once a week, repeated during ten or twelve weeks, or else sublimate injections of  $\frac{1}{6}$  of a grain (0.01 gram), repeated every other day until ten, twelve, fifteen, or twenty have been given, according to the severity of the case. A tablet (suggested by the late Dr. Jones) containing  $\frac{1}{6}$  of a grain (0.01 gram) of bichloride of mercury,  $\frac{1}{4}$  of a grain (0.016 gram) of sodium chloride, and  $\frac{1}{16}$  of a grain (0.004 gram) of muriate of cocaine, has been found particularly useful. In spite of the well-known germicidal properties of sublimate, it has been the writer's custom to dissolve this tablet in a sufficient quantity of water and to boil it in a test-tube before making a deep intramuscular injection. It has been the writer's practice never to give iodides and mercurials at the same time, in order to avoid the formation of insoluble salts. The writer has reason to be thoroughly satisfied with the use of the sodium iodide in saturated solution, beginning with ten drops three times a day, well diluted, and increasing the daily dose by one drop until thirty to forty or fifty drops three times a day are reached. Beyond this it is hardly necessary to go, and the writer does not feel warranted any longer

in supporting the claim that satisfactory results in many cases cannot be obtained unless very large doses, such as 150 grains three times a day, are administered. In the treatment of syphilis of the nervous system the writer has abandoned entirely the so-called mixed treatment.

The question has often been raised whether antisiphilitic treatment is of any distinct benefit in *tabes dorsalis*. One cannot claim that the disease has ever been cured. The pupillary reflexes do not return to normal, nor is an absent knee-jerk restored, but there is no doubt that a condition which seemed to be rapidly progressive has been arrested, that patients who have been tormented by severe shooting pains have suffered less, that vesical and rectal insufficiency has been lessened, and in not a few instances impending mental trouble has been averted. The writer has no hesitation, therefore, in favoring the energetic use of salvarsan, combined with mercurials in the initial treatment of *tabes dorsalis*, and of urging that these rather than iodides be given.

In addition to this specific treatment much else can be done. The patient should be induced to lead a life free of all excitement and excesses. Alcohol and tobacco and every other form of indulgence should be restricted to very narrow limits. Physical exercise should be maintained within sensible limits so long as it is at all possible for the *tabic* patient to get about unaided. In view of Edinger's theories, fatigue should be avoided, but the ordinary *tabic* patient must be encouraged to keep his muscles in good condition and to use them in the best possible way so long as it is convenient for him to do so. The *tabic* patient who lies down, as it were, under his disease, seems to succumb much more rapidly than he who endeavors to make the best of it. Among general measures, hydrotherapeutic procedures have met with great favor. While hot baths should be avoided, the tepid full bath with slightly cooler ablutions can be safely recommended. Cold bathing, particularly sea bathing, is not tolerated well, although among our better classes there are many *tabic* patients who feel thoroughly comfortable while continuing the cold water habit, and so long as they do not complain of increasing neuralgic pains it need not be discouraged. This is a point which may well be determined according to the needs of the individual.

Galvanism of the spine and general faradization are still employed. Many a patient feels comforted if not distinctly improved by such treatment, and the improvement that is attributed to mild faradization can be explained by the effect which it has upon the peripheral sensory neurones. General massage, particularly in connection with hydropathic procedures and with electrical treatment, is of distinct benefit. But there is no doubt that more has been accomplished in the successful treatment of *tabic* patients during the last decades by systematic exercise than in any other way. To Frenkel,<sup>1</sup> of Heiden, belongs the credit of having developed a system of exercises so designed as to reëducate the ataxic muscles. It has been clearly shown that with the exception, perhaps, of the most advanced cases, patients who are so ataxic as not to be able to stand or walk unassisted can regain the use of their muscles and can be

improved to such an extent that they can follow their ordinary occupations. Those who are specially interested in the details of these exercises can find full particulars in the monographs of Frenkel and Goldscheider,<sup>1</sup> also in the writings of O. Foerster.<sup>2</sup> Maloney<sup>3</sup> has studied the relation of fear to ataxia and has elaborated a series of educational exercises based upon this doctrine tending to eliminate the purely psychologic factors of ataxia.

In addition to the general measures, purely symptomatic treatment must often be used. So far as the pains are concerned, nitrate of silver has been endorsed by many, but its good effects are questionable. The use of pyramidon or aspirin, in combination with codeine, is sometimes of value. A distinct caution should be given regarding the use of opiates, particularly the use of hypodermic injections of morphine. Many a tabic patient has also become a marked morphine habitué and has thus made conditions worse than they would otherwise have been.

The treatment of gastric crises calls for good judgment. It is best to give antineuralgic remedies, but during the crises the stomach and bowels should have complete rest, hot applications should be applied over the abdomen, the patient kept in a recumbent posture, and, if drugs must be given, a combination of cerium oxalate and codeine may be used. Rodari has advised the use of pantopon with atropine. In not a few cases special attention has to be directed to the condition of the bladder. Cystitis, with all its attendant dangers, is a common occurrence. This is to be treated according to well-received principles, and particularly in those cases with considerable residual urine, the bladder should be emptied at regular intervals and washed with mild antiseptic solutions. There is danger in allowing the patient himself or even a nurse, unless he be specially trained, to carry out these procedures. It means much for the comfort of the average tabic patient if his cystitis is held in check. In spite of all precautions there is greater danger to the patient from his cystitis than from almost any other condition. The constant use, for a prolonged period of time, of hexamethylenamine need only be mentioned.

If one were to enumerate every possible form of symptomatic treatment, it would be necessary to run the gamut of almost every drug in the Pharmacopœia. A word should be said regarding the attempt to influence a rapidly progressive optic nerve atrophy by specific treatment. From a large experience it is difficult to claim that amaurosis has been completely averted in any one instance. At the same time the claim can be put forth that the development of total blindness seems to have been postponed for a considerable period by the discrete use of antisymphilitic measures. It is, however, in these very few cases that one should be most careful not to push the measures to the extreme, although the writer is not able to endorse the opinions of some that the progress of optic nerve atrophy is made more rapid by the use of these measures. It is wise, however, to have the vision tested carefully during the treatment in

<sup>1</sup> *Anleitung zur Hebungs Behandlung*, etc., second edition, Leipzig, 1904.

<sup>2</sup> *Physiologie u. Pathologie der Coördination*, Jena, 1902.

<sup>3</sup> *Jour. Nerv. and Ment. Dis.*, 1913, xl, 681.

any cases of amaurotic tabes, and if the vision is growing rapidly worse to desist from this therapeutic plan.

Quite as important as, if not more important than, drug treatment, is the psychic treatment. No patients are more liable to general despondency, to marked hypochondriasis, than those suffering from locomotor ataxia. They should be given every possible consideration, be encouraged in every way, and be told that in spite of having an incurable disease, the symptoms may remain stationary for a long period, and that life can be made tolerably comfortable in spite of the disease.

### GENERAL PARESIS.

General paresis of the insane, or dementia paralytica, represents one of the severest forms of nervous disease, and is characterized by a long series of physical and mental symptoms. From insidious beginnings the disease progresses, during a period of several years, to a condition of complete dementia. The majority of cases occur in persons between the ages of thirty and fifty, although its occurrence is not unknown in children (juvenile general paresis) and also in persons entering upon the senile period. In the former there is generally a history of hereditary lues, while in the latter, syphilitic infection has been acquired late in life.

**Etiology.**—General paresis is a disease of civilization, although civilization alone can hardly be held responsible. Krafft-Ebing's dictum that it is due to "civilization and syphilization" has hit the nail on the head. The Chinese, Turks, and the Irish were supposed to be tolerably free of the disease. The same was at one time supposed to be true of the negro race, but in this country we have had ample opportunity to discover that neither the Irish nor the negroes enjoy immunity. It is well established that in only 40 or 50 per cent. of the patients is there any family history of mental or nervous disturbances, of dipsomania, or of any other serious form of degenerative nervous disturbance.

The disease affects men very much more frequently than women, the proportion, according to some, being nearly 7 to 1, according to others, 2 to 1. Among 70 cases in my private practice, 5 were women (7.1 per cent.). In all large cities general paresis, and particularly general paresis in the female, is far more frequent than in the country. Among men the disease is supposed to be commoner among the well-to-do and among those who have indulged in all sorts of excesses. The clergy are known to be relatively free from the disease, while the German statistics show that army officers yield an unusually large percentage. Women of the lower classes are distinctly more frequently the victims than are those of the better classes. These facts point to the well-established conviction that the chief etiological factor of general paresis is syphilitic infection. For many years we have had to be content with statistical proof, and those of us who have had a large experience have regarded it as almost axiomatic that the man who presents symptoms of general paresis must also present some symptoms of constitutional syphilis. In the writer's series in private practice there have not been more than two or three in whom there was



any doubt as to the preceding specific infection. Of late years we have been made independent of statistical proof. The examination of the cerebrospinal fluid has shown a pleocytosis which occurs only in syphilis or in a disease of syphilitic origin. Moreover, the Wassermann reaction has shown an unusually high percentage of positive reactions in the blood of general paretics. Some of the figures have been above 85 and none much below 70 per cent. In 31 cases of the writer's the reaction was positive in only 67.7 per cent. (previous treatment may have influenced this). The discovery of the presence of the *Spirochæta pallida* in the cortical tissue of paretics has removed every possible doubt. The findings of Noguchi and Moore have been corroborated by many others. There may be special exciting causes, but without syphilis general paresis does not occur. The frequent occurrence of general paresis in husband and wife teaches the same lesson.

A period varying from five to twenty years or more may elapse between the initial infection and the first development of the disease. It is doubtful whether thorough or negligent treatment shortly after the initial infection has much influence upon the development of general paresis, but the mere fact that this is still a matter of doubt makes it incumbent upon every physician not only to treat the initial symptoms of syphilis, but to subject every person who has been infected with syphilis to repeated courses of treatment, whether such person presents symptoms or not. With the Wassermann reaction and the spinal cell count as guides there is no sufficient reason why any patient should be allowed to take his chances. The least the physician can do is to demand that the patient have an examination of the blood made at least twice a year, and if there be any positive reaction, then surely a course of treatment is necessary, whether that patient present any specific symptoms or not.

Given a syphilitic infection, any excesses, particularly alcoholic and venereal, may play the part of further exciting causes. Overwork and worry have been assigned as additional etiological factors. Worry and emotional excitement play an important part in the development of many psychic conditions, and may have a bearing upon the development of general paresis, but overwork alone, and more particularly overwork without excesses, is rarely followed by any serious results. Excesses are harmful; work, and even overwork, as a rule, beneficial. Of the many additional exciting causes, the author feels that trauma of the skull is the one which deserves greatest consideration. Among the laboring classes several cases have been seen, in which the first symptoms of general paresis appeared within a few weeks following serious head injury, but in not one of these cases could the additional factor of a syphilitic infection be excluded. Ziehen is of the opinion that the syphilitic virus is apt to exhibit itself in the brain after its resistance to disease has been diminished by severe trauma. Possibly the shoe fits the other foot, and it would be safer to say that syphilis reduces the resistance of the brain and makes the effect of an injury more evident than it otherwise would have been. There seems little doubt that alcoholism is an etiological factor of some importance. When it is able to exhibit its effects upon the system previously weakened by syphilis, the damage it does is doubly great.

**Pathology.**—The morbid changes prove it to be a disease not merely of the brain, but of the spinal cord as well. The popular designation of the disease as “softening of the brain” is wrong in every particular. Macroscopically we may note thickening of the skull, with loss of the diploë, adhesions between the dura and the skull, a pachymeningitis, a condition of external and internal hydrocephalus, a cloudy and thickened appearance of the pia, particularly over the frontal portion of the brain; adhesions between the pia and cortex, a granular ependymitis, and, above all, a very marked atrophy of the cortex. The fissures appear unusually deep and the gyri small. Arteriosclerotic changes are noticeable, more especially in the basilar arteries. The weight of the brain is considerably reduced, atrophy of the frontal and parietal lobes being responsible for this reduction, whereas the temporal and occipital portions of the brain are more nearly normal. On section of the cortex the gray matter appears very considerably reduced in volume.

There has been considerable discussion as to whether or not one could recognize general paresis on the postmortem table. From macroscopic appearances alone this would hardly be safe, although we could make a fortunate guess in perhaps 50 per cent. of the cases. The researches of Cramer, Binswanger, and Nissl and Alzheimer, have thrown a flood of light on the histological changes. The pia is invariably diseased, with distinct infiltration with plasma cells and lymphocytes. All the blood-vessels show either proliferating or degenerative processes. The cortical substance itself contains diseased bloodvessels. There is a proliferation of the endothelium, the elastic tissue is increased, the adventitia proliferates, and the adventitial lymph spaces are dilated and infiltrated. As the disease progresses the bloodvessels undergo regenerative changes. The ganglion cells suffer the greatest change. Many of them undergo complete atrophy, and those that are retained and at all recognizable exhibit every variety in change of the cell architecture. The changes in the interstitial tissue and in the bloodvessels are so great that the question has arisen whether the cells are primarily or secondarily affected. At all events, it is certain that in general paresis the nuclei and the neuroglia fibres are increased, that there is a proliferation of spindle cells, and that these are associated with marked changes in the bloodvessels, some claiming that the entire process may be conceived as a neuroparalytic hyperemia ending in a secondary lymph stasis, and that this lymph stasis is responsible for the degeneration of the nerve elements.

MacIntosh and Fildes<sup>1</sup> state that the lesion of dementia paralytica is as typically syphilitic as a lesion of the liver. It occurs as an inflammatory reaction about the *Spirochæta pallida* situated among the nerve cells. A reaction of the parenchyma rapidly leads to degeneration, etc. According to these authors dementia paralytica is a tertiary syphilitic parenchymatous encephalitis.

In spite of recent advances, it is well to hark back to the important fact that Tuzek first insisted upon—that the tangential fibres are destroyed in general paresis. It was supposed by some that these fibres disappeared,

<sup>1</sup> *Brain*, 1914, xxxvii, 178.

particularly in the association areas of Flechsig. Others maintain that the atrophy is diffuse over the entire cortex. While each one of the pathological features of general paresis may occur in other conditions, it would seem fair to conclude that the disappearance of the tangential fibres, proliferation of the glia, marked vascular infiltration, together with the atrophy of the gyri, particularly in the frontal and parietal lobes, would enable one to recognize general paresis. It should also be stated that other portions of the brain do not escape; degenerative changes are to be found in the basal ganglia, particularly in the optic thalamus. Westphal, Siemerling, and others have found degenerative changes in the nuclei of the ocular muscles, and such changes have also been noticed in the nuclei of the pons and the medulla oblongata. Westphal was among the first to note a degeneration of the various systems in the spinal cord. The pyramidal tracts and the posterior columns are those most frequently affected. The degeneration of the posterior columns has been held by many to be identical with the changes in the *tabes dorsalis*; others have thought it to be distinct therefrom. The frequent association of *tabes* with general paresis may be more readily explained if we presume these changes to be identical in both diseases. Degenerative changes have also been found in the cerebellum and even in the peripheral nerves. The student who wishes to investigate this special part of the subject is referred to the articles of Nissl and Alzheimer,<sup>1</sup> Cramer,<sup>2</sup> Joffroy et Leri,<sup>3</sup> MacIntosh and Fildes and Nonne.<sup>4</sup>

**Symptoms.**—There are few diseases so easily recognized as general paresis when the symptoms are well developed; there is, on the other hand, no disease of the central nervous system which begins so insidiously and which is diagnosed so often on mere suspicion. If once suspected, in nine cases out of ten the suspicion is corroborated by the further progress. The disease is far reaching, not only for the effect that it has upon the patient himself, but often enough it has important bearings upon business and family ties. Nine times out of ten the patient who is brought to the physician's office with a blank countenance, who enters the room with an air approaching indifference, or at times in a state of excitement, who by the first word he utters reveals the tremor of speech, whose attitude shows that he fails to appreciate the importance of a medical examination, may be recognized as a sufferer from parietic dementia. The blank stare, the parietic speech, and the indifferent manner are the first conspicuous signs. The disease is characterized chiefly by a progressive form of dementia, by a tremulous speech, by epileptiform and apoplectiform attacks, and by disturbances in the pupillary reaction.

The symptoms may be divided into psychical and physical. The *mental* symptoms are much more important than the physical. Many a patient is aware of a change coming over him; others do not appreciate that they are in any wise different from their former selves, and resent the suggestion of illness. In the earliest stage the patient may have what appear to be ordinary neurasthenic symptoms, such as mild depression,

<sup>1</sup> *Histol. und histopathologische Arbeiten von Nissl*, Band i, 1904.

<sup>2</sup> *Handb. der path. Anat. des Nervensystem*, Band ii, chap. 41.

<sup>3</sup> *L'Encéphale*, 1908, No. 4, p. 322.

<sup>4</sup> *Deutsch. Zeitschr. Nervenhe.*, 1913, xlv.

a feeling of pressure on the top of the head, irritability, sleeplessness, vertigo, and easy fatigue. In association with these a change is noticed in the patient's emotions and general behavior. The man who has been extremely punctilious about business engagements neglects them and can give no special reason why he has neglected them; in fact, he fails often to see the importance of having done so. The patient who has practised a rational economy becomes wasteful; the man who has been sober in his dress takes to the wearing of garish colors; the family man who has been respectful toward his wife and children, toward his employees and his servants, shows an entire lack of respect, and instead of the decent treatment which he was accustomed to accord them, behaves tyrannically, if not brutally. Such changes denote a marked defect of judgment as to the relations between the patient and his fellow-beings. The same patient will squander money, sign checks recklessly, grow irritable over small trifles in business, neglecting larger and more important duties, thus proving that his judgment has become defective. Impaired judgment may lead the patient into criminal acts. Petty thefts, indecent exposures, improper relations toward subordinates, are some of the acts committed in the earlier stages of general paresis.

With this defective judgment *defective memory* is associated at an early date, so that the patient forgets the facts that everyone is expected to know—the day of the week, the month, the year, the name of the President, even the names of his family. The parietic at an early stage may be unable, offhand, to give the names of his children, or of his partner; his memory for recent events is often more impaired than his memory for things that have happened long since. It is curious to see how easily the physician may get at the evidence of impaired memory. As a rule, it is best to begin by questioning the patient as to his age. If he states, for instance, that he is fifty-two years of age, and you ask him the year of his birth, he is unable to give it; ask him how long he is married, he fails to state it correctly; if the wife in his presence supplies the answer, he is unable to state the year of his marriage. A simple sum of subtraction is quite beyond his mental powers. If asked to write his name and address, he will be able to do it correctly in the majority of instances, unless he is in a very advanced stage. Ask him to write the names of some cities which have not come within his daily correspondence, and you will soon detect his inability to do so. The things which he has known most thoroughly he may be able to do; those which require a little reflection he fails in. In the early stage the patient may write New York, Boston, and Buffalo correctly, he is apt to fail on Philadelphia, Rochester, and Constantinople, which last is the stumbling block for many. In every instance it is well to assure one's self that the patient spelled correctly before the disease came on. His memory may also be tested by putting before him the simplest sums in arithmetic. The ordinary additions he may be able to do, but ask him to multiply 8 by 13 or 7 by 8, or ask him to divide 120 by 15, and one quickly discovers the limitations of his knowledge. Far from being annoyed at his mistakes, the average parietic shows either an indifference to the test or considers it a huge joke.

The difference in the response on the part of the neurasthenic and the

paretic is so marked in his answer to these simple tests, that whatever suspicions one may have entertained as to a mere neurasthenia are soon set aside by the results of this short examination. The outcome of these tests is that the first signs of dementia are quickly discovered, and as the disease progresses the dementia becomes more marked, so that finally the patient has forgotten the most elementary facts and his memory will grow so feeble that in the course of time he is nothing but a vegetating organism. So much for the rapidly progressive dementia.

Associated with this dementia are other psychic symptoms. We have alluded to unusual irritability out of all proportion to the provoking cause. Of great import are the *delusions of grandeur*, which were by many considered almost pathognomonic, but which seem to many of us to play a less important part in the diagnosis than they did fifteen or twenty years ago. What the cause of these changes in the general clinical picture may be cannot be discussed at this point, but every now and then we still hear patients give expression to their exalted state of feelings, and the point that is characteristic of the delusions of grandeur of general paresis as compared with the delusions of grandeur of paranoia, is that in the former the delusions are extravagant, beyond all reason, and wholly unsystematized. They are the transitory and fleeting expressions of the moment, or mere extravagant utterances, and do not constitute the basis for purposive actions as they do in paranoia.

The delusions of grandeur of the paretic are in general coincident with the euphoria, and vary with the station in life which he occupies. The paretic laborer may exhibit his exalted state of mind by travelling about, as one poor fellow did, all day long on trolley cars to prove that he owned the company. The merchant in the earlier stages of paresis may imagine himself doing millions instead of thousands of business per year. The stock broker imagines himself commissioned to corner steel or some other stocks. Unlike the paranoiac, he does not exact from others the respect and the homage due to his position.

In relatively few cases the delusions are of a hypochondriacal and even of a melancholy tinge. The belief of an entire absence of stomach or bladder, the idea that the food instead of being digested passes into the head and not into the stomach, or the notion of a change in the genital organs, these and other equally absurd delusions occur in general paresis. Maniacal and deeply melancholic conditions, states of extreme anxiety and fear, also constitute a peculiar feature of the psychical condition. The true value of these is easily recognized, first of all by their absurdity and by the association of these delusions with some one of the other physical symptoms characteristic of general paresis or with the distinct evidence of a rapidly developing dementia.

It is worth noting that in the *early stages* many patients realize the impending trouble. They may be conscious of the lapses of memory or of the difficulties in transacting ordinary business, and some notice the difficulty of speech. The writer has had a number of patients who made the diagnosis of general paresis on themselves, and in whom the suspicion has unfortunately come true. This was particularly striking in the case of a young lawyer, who had lost a brother from general paresis some years

previously, and who had a distinct recollection of the symptoms as the brother exhibited them; at the time he consulted me he recognized that he himself was exhibiting the symptoms of the parietic brother. After the initial stages have passed the patient soon lapses into a condition of apathy, and many of them become entirely oblivious to everything going on around them and are bedridden until released by death.

The *physical signs* are quite as helpful in establishing the early diagnosis as are the mental symptoms. Reflex immobility of the pupils is one of the earliest symptoms, and often precedes the onset of other signs by many years. In some patients we have the typical Argyll-Robertson pupil, in others there is an immobility both to light and during accommodation. Irregularity in the contour of the pupil, either when the pupil is dilated and particularly when contracted, together with complete immobility, suggest an active specific virus. Inequality of the pupils and paralyzes of the external ocular muscles also occur, the latter more frequently in the cases that are associated with tabic symptoms.

*Speech disturbances* have an unusual significance and the suspicion of general paresis rests more often upon this one symptom than upon any other. The disturbance consists in a tremor of speech and in the inability to pronounce words with labials and consonants, particularly with *l* and *r*, and in the inability to repeat test sentences consisting of words joined together rather by sound than by their inherent meaning. Ask the parietic to say "truly rural," "artillery cavalry brigade;" ask him to repeat such sentences as "Around a rugged rock a ragged rascal ran;" and you will find not only difficulty in articulation, but also a memory which is so defective that the patient forgets some of the words or some of the syllables and makes a jumble of the entire sentence. Neurasthenic and other non-parietic patients may hesitate and make some few mistakes when asked to repeat these test sentences, but on the second or third trial they correct their mistakes and pronounce the sentences well; above all, they show no such defective memory as the parietic patient does.

In connection with the speech disturbance, *tremors* of the facial muscles, of the tongue, and of the hand are particularly noteworthy. The tremor of the fascial muscles is, perhaps, the most significant. When the parietic is asked to show his teeth, the entire face, including the lips, quivers; when asked to protrude the tongue, he exaggerates the movement, the tongue is jerked to and fro; and in most characteristic fashion the parietic, instead of pushing out the tongue once or twice, as the ordinary patient would do, keeps on doing it until he is told to stop. The facial tremor has a further significance, inasmuch as it is seen practically in no other condition except in acute and chronic alcoholism. The existence of this tremor alone necessitates the differential diagnosis between general paresis and alcoholism. The tremor of the hands is of a coarse kind and is more an awkwardness of movement than anything else. This suggests another disturbance so common in the general parietic, viz., a general awkwardness of movement associated with the tremor. In tying the cravat, in buttoning or unbuttoning a vest, in fastening suspenders; in the case of women, in the attempt to sew on a button, to hem a handkerchief, even to fasten a hairpin or a breastpin; in short, in attempting to

do the most accustomed acts of every-day life, the patient fails to do these successfully, and spends an unusual amount of time in accomplishing them. He fumbles and grows excited over the most trifling actions. The same tremor and awkwardness are shown in the attempt to write. The paretic may retain the power to write his name and the words which he has been accustomed to write again and again. Oftener his writing is tremulous, his memory is so defective that he omits letters and syllables in the most ordinary words, and as the disease advances he forgets even to write his name. The difficulties in writing may occur so early that the patient is startled at his inability, and worries about this more than about any other symptom.

The *deep reflexes* may be either lost or exaggerated. In keeping with the changes which are known to occur in the spinal cord, it is not astonishing that this difference should occur. In some we have, together with the immobility of the pupils, a loss of the deep reflexes, of the knee-jerks, and of the Achilles tendon reflexes—the tabic series of symptoms. In others the reflexes are exaggerated, the knee-jerks are very much increased, and ankle clonus may be present. The exaggeration of the reflexes must be considered, particularly in those cases in which there is a question as to the condition being one of neurasthenia or of general paresis, although in neurasthenia it is most unusual to have an ankle clonus.

A general *muscular weakness* is also frequent, particularly in the second half of the disease, and offers excuse for the term *dementia paralytica*, which, in perhaps more than one-half of the cases, would seem to be unwarranted. With this muscular weakness, particularly if the patients are kept in bed, a general flaccid, atrophic condition of the muscles is apt to be associated, due more to disuse than to anything else.

Among the physical symptoms we must also include convulsive and apoplectiform *seizures*, which occur during the progress of the average case. While the rule holds good that these seizures occur in the second half of the disease, they sometimes constitute one of the earliest signs. The writer has always insisted that if a patient past middle life, who has neither cardiac nor renal disease, develops epilepsy or has an apoplectic attack, the suspicion is justified either of active syphilis or of general paresis, if not of both.

Not infrequently the neurologist is asked to see a patient who is supposed to be in a uremic convulsion. Examination of the urine, however, fails to show any trace of kidney affection, and the suggestion that the convulsion may be the first symptom or an early symptom of general paresis comes as a surprise to the general practitioner. In the writer's experience, patients who exhibit apoplectiform or convulsive seizures at an early period run a very rapid course, the disease often ending fatally within six months or a year after such convulsions or apoplectic attack. The convulsive attack is of the ordinary epileptic character, and can in no wise be distinguished from the typical seizure, unless it be that the patient recovers very rapidly and appears, possibly for a week or more thereafter, to be in unusually good health. A little later, however, the first signs of the dementia set in, and the patient is found to present other characteristic signs of general paresis. The apoplectic seizure may result in a

temporary hemiplegia, from which the patient also may recover with unusual rapidity, and in other patients there may be, with or without hemiplegia, a transitory aphasia. The symptoms of these apoplectic or apoplectiform seizures disappear with unusual rapidity, but they leave the patient, as a rule, in a disturbed state, and with every recurrent attack—some paretics have six or even more such seizures—the mental condition becomes more and more apathetic and there is a steady deterioration. The rapid disappearance of the apoplectic symptoms leads to the suspicion that these attacks are not due to the ordinary vascular accidents, but that they must be due to some temporary interference with the circulation. In patients who have died of general paresis after apoplectic seizures no sufficient cause has been found for the apoplexy.

*Vasomotor* and *trophic* disturbances are not unknown. Among these we need merely enumerate bedsores, perforating ulcer of the foot, arthropathies, gangrene. There is also at times an unusual fragility of the bones, so that fractures occur most easily and not infrequently lead to the suspicion or to the accusation that the patient has been severely handled by an attendant. In asylums hematoma of the ear is looked upon with especial suspicion as a symptom of unusually grave import. It occurs not only in general paresis, but also in many other psychic disorders. While it may be due to slight trauma, it surely would not occur if it were not for the marked tendency to trophic disorder. Pruritus, salivation, and universal cyanosis occur in rare instances, but have no pathognomonic value.

**General Course.**—Enough has been said to indicate that the disease begins in insidious fashion, that there is often a suspicion of serious disease impending before the definite diagnosis can be made, and that in the majority of instances, if the suspicion has once been entertained, the worst fears, unfortunately, prove to be well founded. After the initial period of excitement or depression, accompanied either by hypochondriacal delusions or delusions of grandeur, after the development of the typical mental and physical symptoms, the disease passes into the stage of progressive dementia. After the lapse of months or of one or two years, during which time the patient has been in a state of euphoria, and, as a rule, is the only one in the entire circle ignorant of the severity of the symptoms, he passes into the terminal stage, in which he is bedridden, a mere vegetating organism, soiling himself, developing bedsores, becoming more and more emaciated; he dies from exhaustion, an apoplectic seizure, or intercurrent diseases.

**Duration.**—It is commonly supposed that the disease runs its fatal course in two, three, or five years, but every alienist has seen cases that have run a course of ten, fifteen, or even twenty years. Many of us have obtained the impression that general paresis does not run as rapid a course as seemed to have been the rule years ago. It is difficult to decide whether this is due, on the one hand, to earlier recognition, so that the period of observation is longer, or to the more careful and more persistent antisiphilitic treatment which these patients now receive. But that careful treatment has much to do with the prolongation of life in general paresis is unquestionably true. Some of the longest-lived parietic patients



are among those who have been nursed at home, where greater care can be given them than in the average asylum. There is also a further possibility that some of these long-lived cases of general paresis are, after all, cases of syphilitic pseudo-paresis, and in this connection it is necessary to call attention to the marked remissions occurring in the disease. Two and three such periods of remissions amounting to months and even years are not unknown. The most troublesome remission, however, is the one which frequently sets in after the disease was first suspected and after a proper course of treatment has been instituted; the improvement may be so great that doubt arises as to the diagnosis. This doubt is soon dispelled by the development or continuance of the typical symptoms.

**Prognosis.**—This is most unfavorable if we allow for the few questionable cases which are supposed to have recovered. It is safe to predict an early fatal termination in almost every case of general paresis, and yet the experience of each alienist will undoubtedly bear out that of the present writer, who, in spite of his knowledge of the grave prognosis, has insisted that recovery is not altogether impossible, or at least that such pronounced remissions may occur that the patient may be able to resume his duties and be a useful member of his family and of society for a number of years. This is supported by the fact that several patients in whom, more than twenty years ago, at least three and four prominent alienists made the diagnosis of general paresis are at this day mentally well, although they still present some of the physical signs of the disease. There are disorders in which the diagnosis of general paresis is made for the want of better knowledge, and until we learn to discriminate better between the various forms of general paresis, it is well to know that some cases may behave contrary to the ordinary rule.

**Diagnosis.**—A disease presenting as manifold a series of symptoms will naturally give rise to great difficulties in diagnosis. When the mental condition is in doubt the presence of the physical signs is of the greatest possible import. Thus, if a patient presents what would ordinarily be supposed to be symptoms of neurasthenia, and if he, in addition, has small pupils and either diminished or exaggerated reflexes and a slight peculiarity of speech, the probability of his having more than a mere neurasthenia becomes very great. In attempting to differentiate between neurasthenia and general paresis stress must be laid on the fact that while the neurasthenic may complain of lack of concentration, of his inability to recall things, of pains in the head resulting from the effort of thinking, etc., he presents no signs of actual mental deterioration. Yet the distinction cannot always easily be drawn. In the writer's experience a patient was suspected of suffering from neurasthenia until the occurrence of an epileptic attack helped to clinch the diagnosis of general paresis. Parietic speech can hardly be mistaken for any other speech disturbance, but the hypochondriac and the neurasthenic are apt to notice slight hesitancy of speech which may occur within normal bounds, and noticing these may accentuate them; but even if there is slight hesitancy in articulation, it is very rare to have any such evidence of defective memory associated with defective speech as occurs in general

paresis. Moreover, the neurasthenic is much more conscious of his disturbed condition, and is much more likely to attach importance to it than the parietic does. Marked as the difference between the two diseases is, every now and then cases will occur, particularly in those who have been known to have had syphilis, in which for a period of a few weeks or even longer there may be a doubt as to whether neurasthenia or general paresis is in process of development. The examination of the cerebrospinal fluid and the Wassermann reaction of the blood will give further help. The Wassermann reaction of the cerebrospinal fluid is positive in almost every case of general paresis.

The occurrence of epileptiform and apoplectic attacks is of great importance in doubtful cases; also when the question arises whether the case is one of general paresis or of multiple cerebrospinal sclerosis. The writer has seen few cases in which there was any reason to hesitate with reference to *disseminated sclerosis*. The tremor, the scanning speech, the marked exaggeration of all the deep reflexes, the nystagmus, are symptoms which are never or rarely as marked in general paresis as in disseminated sclerosis, nor does the mental condition accompanying multiple sclerosis at all suggest dementia paralytica. A difficulty may occasionally arise if one sees a patient with disseminated sclerosis in or near the terminal stage. In that case, without previous history, a condition of dementia paralytica might be suspected, but the protracted course, the existence of physical signs before the mental symptoms developed, and the entire aspect will give the diagnosis.

Far more difficulty is experienced in attempting to differentiate between general paresis and *syphilitic pseudo-paresis*, or lues cerebri. The difficulties are increased by the fact that the two conditions are very similar, and that even the pathologist cannot easily establish the line of demarcation. Clinically it may be said that in lues cerebri there is always complete immobility of the pupils both to light and during accommodation. The onset is often preceded by other specific accidents, as previous paraplegias or hemiplegias that have ended in more or less recovery. The dementia is not as rapidly progressive as in the genuine form of dementia paralytica. Above all, we may argue the presence of a lues cerebri from the effect of antisiphilitic treatment. This is never, or rarely, as prompt in general paresis as it is in this special class of cases. After all has been said with regard to this differential diagnosis, the fact remains that cerebral syphilis often gives rise to a combination of symptoms which it is impossible to differentiate from genuine dementia paralytica. Furthermore, everyone has seen cases which have been diagnosed as syphilitic pseudo-paresis, pass into genuine forms of the disease, so that it has been the writer's habit, whenever he has been compelled to make the diagnosis of a pseudo-paresis syphilitica or of brain syphilis with mental symptoms, to state that the case is more hopeful regarding the possible effects of treatment than a typical general paresis would be, but that these specific forms often terminate in typical genuine paresis.

There is also an *alcoholic pseudo-paresis* which may lead to some confusion but the difficulties are not nearly as great as in the case of syphilitic disease simulating general paresis. The psychic symptoms

due to alcoholism may result in a demented condition, but, as a rule, there has been a history of delirium tremens, there are very marked delusions and hallucinations, and, more than this, there are the distinct physical symptoms of alcoholism, and the pupillary reflexes are rarely as much involved as they are in syphilis or in general paresis.

There is also an *arteriosclerotic dementia* which we see in older persons, which may have some resemblance to general paresis, but, first of all, the age of the patient argues rather in favor of an arteriosclerotic process than general paresis; the dementia is not as rapid and rarely as complete; and, as a rule, before any suspicious mental condition is reached there has been a history of hemiplegia, transitory attacks of aphasia, vertigo, and of numerous other conditions associated with arteriosclerosis.

After giving due consideration to all these points the fact remains that when general paresis is suspected it is present in fully 90 per cent. of the cases, and that in the remaining 10 per cent. it will be most difficult to state whether or not the diagnosis of general paresis or of some forms simulating general paresis must be maintained.

**Treatment.**—Granted that in the vast majority of instances general paresis is an incurable disease, there is, nevertheless, much to do if the patient is to be properly cared for. In the initial stages the proper treatment may bring about a cessation of the symptoms or a very marked remission, and the patient may be enabled to return to his family and to his work for an indefinite period. The first essential is that the patient be kept free of all worry and excitement, and that he be subjected to as few new mental impressions as possible. To send a paretic travelling with an attendant for a change of environment is a grievous error. The paretic brain needs rest above all things. Many writers insist that all paretics shall be committed at once to institutions for the insane. It is well to do this, in view of the fact that the paretic is wholly irresponsible and may suffer bodily harm and inflict injury to others. In the crowded streets of our cities a man in the initial stages of general paresis is scarcely able to take care of himself properly. The paretic surely needs supervision at a very early stage. On the other hand, among the well-to-do classes, if a reliable attendant can be secured, it is often feasible to retain the paretic in his own home very much longer and with far less difficulty than in the case of patients suffering from other forms of mental derangement. Other things being equal, the writer cannot support the contention that every patient suffering from general paresis must at once be confined in an asylum. Each case should be judged upon its individual merits.

The close relationship between syphilitic disease of the brain and general paresis, the rapid development of pseudo-paresis in syphilitic subjects, make it incumbent in every instance to give the patient the benefit of salvarsan treatment with mercurial injections (see page 830).

If the Wassermann reaction of the blood is positive and if the study of the cerebrospinal fluid shows a positive Wassermann reaction and a marked increase of the cells, there is every reason to persist in an intensive course of antisymphilitic treatment. Every neurologist has seen cases in which remarkable remissions have occurred after early

salvarsan treatment. The very fact that with the refined methods in diagnosis the doubtful cases of general paresis seem to be increasing makes it more and more important that active treatment should be administered in every case but the physician must exercise judgment lest the treatment make further inroads upon a constitution that is already wasting away with the disease. To persist in often repeated courses of salvarsan injections simply because the Wassermann reaction remains positive and the cell-count is high is not the wisest procedure, because experience proves that in a very large majority of cases of general paresis salvarsan treatment will not be able to change a positive Wassermann of the blood and of the cerebrospinal fluid to a negative phase, and the cell count may not always be diminished. If salvarsan treatment cannot be given, mercurial treatment with iodides may be attempted.

The *intraspinous* treatment of general paresis with salvarsanized serum, whether according to the method of Swift and Ellis or that of Ogilvie<sup>1</sup> has not yielded satisfactory results so far as the experience of the writer goes. If we remember that the spirochetes in general paresis are imbedded in the tissue of the cortex and away from bloodvessels or lymph spaces, we can understand why this direct application of salvarsan can not be as effective as might be expected on theoretical grounds.

The suggestion has been made to produce artificially an increased pleocytosis in general paresis and thus to counteract the toxin of the disease, basing this on the old experience that remissions in dementia paralytica were very apt to occur after febrile and infectious processes. With this end in view, injections of tuberculin and of neucleinate of soda have been recommended, and while a few writers think they have seen very remarkable remissions, the author's experience after repeated trials has led him to believe that these methods do not accomplish much more than can be effected by judicious intravenous salvarsan treatment. Donath insists that there was a marked improvement in 70 per cent. of the cases treated by injections of nuclein according to the following formula: Nucleinate of soda and chloride of sodium, 2 grammes each, in 100 grammes of sterile distilled water. This quantity to be injected at one or two sittings and to be repeated at intervals of five to seven days.

In every case there is need of purely symptomatic treatment. The ordinary states of excitement may be influenced by the rational use of bromides, codeine, and the hydrobromate of hyoscine (gr.  $\frac{1}{100}$ , by mouth or hypodermically). The insomnia, which is very troublesome in many cases, is to be combated by the use of bromide, chloral, veronal, trional and paraldehyde in the ordinary doses. Mild hydiatic procedures are in order, but extreme forms, such as cold spinal douches and the like, should be avoided. In the chronic and bedridden cases the function of the bladder has to be carefully watched; the tendency to bedsores must be carefully considered. If the patient cannot feed himself, the food must be administered carefully. In the advanced stages treatment is purely symptomatic. The rational medicinal treatment and careful nursing in the earlier stages are often followed by encouraging improvement.

<sup>1</sup> *Jour. Am. Med. Assn.*, 1914, lxiii, 1936.

## CHAPTER XXIII.

### AMAUROTIC FAMILY IDIOCY (TAY-SACHS DISEASE).

By B. SACHS, M.D.

THE name, amaurotic family idiocy, was applied by the writer to a family disease of infancy, characterized by an entire lack of mental development, a progressive weakness of all the muscles of the body, and rapidly developing blindness, with typical changes in the macula lutea. The disease is generally fatal, the patients dying, as a rule, in a condition of marasmus before the end of the second year of life.

In 1881 Waren Tay<sup>1</sup> published an article describing "symmetrical changes in the region of the yellow spot in each eye of an infant." The child was twelve months old. It could not hold up its head or move its limbs; there was weakness but not absolute paralysis of any part. Its entire brain development was slow and poor. At the first examination, the optic disks were apparently healthy, but "in the region of the yellow spot of each eye there was a conspicuous, tolerably diffuse, large, white spot, more or less circular in outline, and showing at its centre a brownish-red, fairly circular spot contrasting strongly with the white spot, and this central spot did not look like a hemorrhage, nor as if due to a pigment, but seemed a gap in the white patch through which one saw healthy structures." Five months later examination showed that the disks had become atrophied, but that the changes in the macula lutea were the same as before. In the same family, according to Tay's reports, three similar cases have occurred, each one of the children presenting ocular symptoms and exhibiting physical conditions that were similar in all respects, and all three dying before the age of two years.

The writer,<sup>2</sup> in 1887, published the history and the postmortem record of a patient who had suffered from a peculiar form of idiocy associated with blindness. The family character of the affection was not evident until a sister of the first patient, born several years later, had become similarly affected. In the course of the next six or seven years the author had occasion to see several instances of the same disease and to obtain the history of a family in which four children had been afflicted with this disease and all had died of it. In 1896<sup>3</sup> the writer was able to give a list of 19 cases, of which 8 had come under personal observation, and by that time the chief symptoms of the disease had impressed themselves so clearly upon his mind that he felt warranted in giving the disease the name Amaurotic Family Idiocy. Since that publication numerous cases have

<sup>1</sup> *Tr. Ophth. Soc. of the United Kingdom*, 1881, i, 55; also 1884, iv.

<sup>2</sup> *Jour. Nerv. and Ment. Dis.*, 1887, xiv, 541; 1892, xvii, 603; and 1896, xxi, 475.

<sup>3</sup> *New York Med. Jour.*, 1896, lxiii, 697.

been described by American and by some European writers. An excellent review of the entire subject, with the complete literature<sup>1</sup> to 1909, and with a careful analytical study of 58 cases, is contained in the work of Wilbrandt and Sanger (*Die Neurologie des Auges*).

**Etiology.**—Up to the present time it has been impossible to assign any definite reason for the development of this disease in special families. In some of those examined by the writer there has been blood relationship between the parents, and in one of the families there had been innumerable psychoses among the relatives of both parents. Injury to the mother during pregnancy has been noted in at least one of the writer's cases. The family predisposition is evident from the fact that 28 cases occurred, to the writer's knowledge, in fifteen families. Carter was the first to call attention to the fact that all of the cases reported had occurred among Hebrews,<sup>2</sup> and even up to the present day not a single indubitable case has occurred among any other people. The racial feature is all the more astonishing because other diseases to which it is closely allied, and above all the juvenile form of amaurotic family idiocy, have been observed and recorded among all races and all nationalities.

It is important to note that syphilis is not an etiological factor. This is of interest not only because other family affections have been attributed to syphilitic infection, but also because the disease bears a distinct resemblance to specific disorders characterized by various forms of dementia and by ocular disturbances (Batten). Moreover, there is a hereditary optic nerve atrophy occurring late in life which is supposed to be due to hereditary syphilis, and with this optic nerve affection the present disease might possibly be confounded.

**Pathology.**—In 1887, before I had fully recognized all the characteristics of this striking affection, I described the condition as one of arrested cortical development and supposed that this was due to what I termed an "agenesis corticalis." I was much impressed at the examination of the brain of the first patient with the confluence of the central and Sylvian fissures and a complete exposure of the island of Reil.<sup>3</sup> The brain was unusually hard, even in the fresh state, and the knife actually grated on removing a small section of the cortex. On microscopic examination the most important changes were found in the cortex, in sections taken from the frontal lobes, the motor areas, from the base of the third convolution, from the first temporal, and from other parts of the cortex. The same changes were found in all the cells. I was struck by the fact that hardly a single pyramidal cell presented anything like a normal appearance. The contour of the cells was either round or elongated, and the cell protoplasm presented every possible change from slight to complete degeneration. In some cells the nucleus and nucleolus were entirely

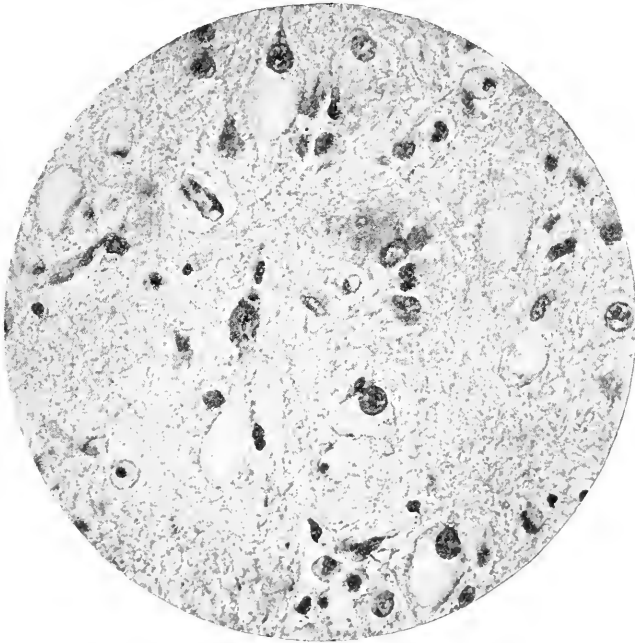
<sup>1</sup> Robert Bing, in the *Ergebnisse der innere Medizin u. Kinderheilkunde*, 1909, iv, 82, has made a careful study of this disease in connection with other family affections; Apert (*Sem. med.*, 1908, p. 25) collected 106 cases; of these, 73 cases occurred in twenty-five families.

<sup>2</sup> Bing is in error in restricting the disease to Polish Hebrews. My first two cases were of German Hebrew extraction.

<sup>3</sup> I do not attach much importance to the confluence of fissures which I described in my first cases.

wanting or were relegated to the margin of the cell. A few years later Hirsch found that the same cellular changes occurred not only in the gray matter of the cortex, but in the gray matter of the entire central nervous system; not only in the cortex of the brain, in the cell ganglia, and in the gray matter of the spinal cord, but even in the spinal ganglia. This was corroborated by a later examination of one of my own cases,<sup>1</sup> and in 1903 I felt warranted in declaring amaurotic family idiocy to be a disease chiefly of the cortex and of the gray matter of the entire central nervous system. These findings have been corroborated by all recent investigators, among whom I wish particularly to mention Spiller, Kingdon, and Schaffer.<sup>2</sup> The last-named writer has devoted himself to the study of the

FIG. 48



Brain cortex cell. Showing degenerating ganglion cells.

pathology of this disease, and has given a most thorough anatomical study of 8 cases. All of his findings have been splendidly corroborated by a recent study of my own.<sup>3</sup> The findings are so characteristic that the diagnosis of amaurotic family idiocy can be made without hesitation from an inspection of the microscopic specimens.

Schaffer pointed out that the chief characteristics are: (1) A widespread cytopathological process; an unusual swelling of the cell protoplasm and of the dendrites; a swelling of the hyaloplasm, which causes a mechanical destruction of the cell fibrils; ultimately the cell body is a mass of

<sup>1</sup> *Jour. Nerv. and Ment. Dis.*, 1903, xxx, p. 1.

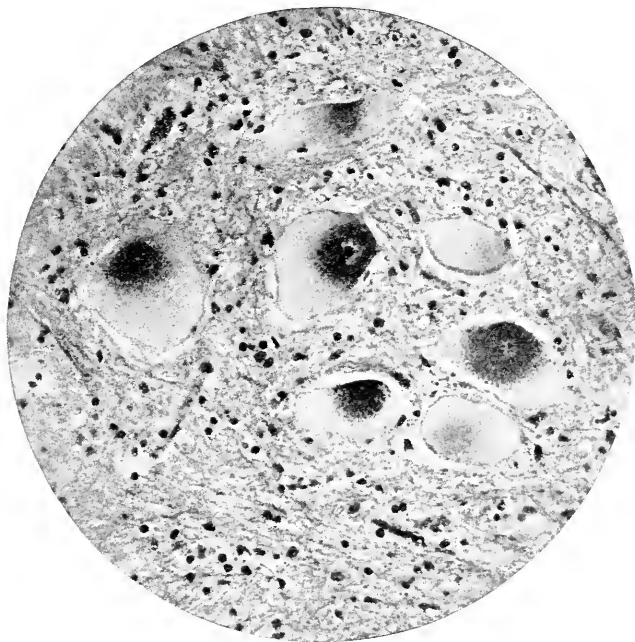
<sup>2</sup> *Zeit. f. die Erforschung. u. Behandlung. d. Jugendl. Schwachsinn.*, 1909.

<sup>3</sup> Sachs and I. Strauss, *Jour. Exper. Med.*, 1910, xii, 685.

detritus. Curiously enough, the axis-cylinder is not involved in this general swelling. (2) Every cell of the central gray matter, both of the brain, the spinal cord, and the spinal ganglia, is similarly affected.

Histologically there is an unusual swelling of the cell body and a more or less complete disintegration of the cell protoplasm. The fibrillæ disappear, so that in many of the cells there are only slight traces left of the endocellular network. The fibrils near the periphery of the cell body are preserved somewhat longer than are those in the central part of the cell mass. The nucleus and nucleolus, if at all preserved, are, as a rule, pushed nearer to the periphery of the cell body, and as the disintegration

FIG. 49



Ganglion cells of the anterior horn of the cord. Showing the excentric nucleus surrounded by the remains of the chromatophilic substance and the marked increase in size due to degeneration of the cytoplasm.

of the mass goes on the nucleus and nucleolus also disappear. A proliferation of the glia, particularly in the cortex, with the development of giant glia cells, has been noted. There is no evidence of any inflammatory process. Changes in the white fibres are unimportant and unessential. It is Schaffer's merit to have laid stress upon the swelling of the dendrites, which appears to be so constant as to be almost pathognomonic. The deterioration of the cell body and the swelling of these cell processes, exclusive of the axone, have been noted in all of Schaffer's and in my own recent case.

Holden, as well as Schumway and Mary Buchanan, made careful histological examinations of the eyes. All these writers agree that the



essential changes are a degeneration of the ganglion cells of the retina and of the nerve fibres of the optic nerves and tracts. They also believe that the white area in the fundus is the result of the swollen and degenerated ganglion cells, which are present in much greater numbers in the macular region than elsewhere, and do not consider this white area to be due to oedema of the tissue as was first supposed.

**Pathogenesis.**—Writers on this disease have been divided into two groups. Some, among whom the writer wishes to class himself, have maintained that amaurotic family idiocy is, in a sense, a congenital disease, even if it is not manifest at the time of birth. The tendency to the

FIG. 50



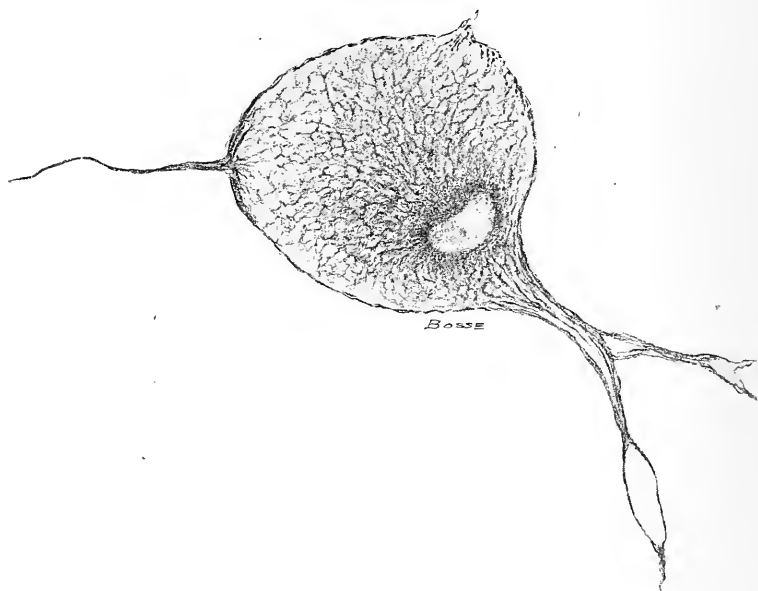
Purkinje cell. Showing the swollen dendrites.

disease is unquestionably born with the child. In the writer's opinion children afflicted with amaurotic family idiocy are possessed of a nervous system so inadequate to the demands imposed upon it that its cells, after having performed their function for a few weeks or months, undergo complete disintegration. In other words, these children have a very limited capacity for normal development. Their central nervous system is not equal to the functions that they are expected to perform for more than the first three or four months of life.

Even Schaffer, who was inclined for a time to class amaurotic family idiocy among acquired diseases, concedes that the entire nervous system of such a child is so delicately organized that it is not able to repair the ordinary physiological waste of its cells. The nerve cells are so easily

exhausted that they undergo a progressive deterioration, which leads to the death of the ganglion cell with a consecutive hyperplasia of the glia substance. These views are so thoroughly in accord with those of the writer that there seems no radical difference between them. Only a few authors now adhere to the view which was held by Hirsch, that the disease is altogether an acquired condition and that it is possibly due to some toxic agent. Hirsch believes that it might be due to some toxic substance in the mother's milk. From the very outset I was loath to accept the explanation of a toxic origin for any profound family disease, and in the case of these amaurotic children Hirsch's view falls to the ground because several of those seen by the writer were not nursed by

FIG. 51



Large anterior horn cell. Showing very well-preserved endocellular network and swelling of dendrites. The fibrils have commenced to disappear in the dendrites. (Bielschowsky.)

their own mothers, but by wet-nurses of a different race and different nationality. However, a child born with a defective nervous system may be more subject to toxic influences than normal children.

**Symptoms.**—The children with this disease have been born at full term and apparently in perfect health. Difficult labor or instrumental delivery has no bearing upon its development. The children do well during the first three to six months of life, when they become listless and apathetic, move their limbs very little, show no interest in their surroundings and show the first signs of visual disturbance which ultimately leads to blindness. As time goes on, the child is unable to hold up its head or to sit up. Its muscles are generally flaccid, rarely spastic. The reflexes may be normal, a trifle subnormal, or somewhat exaggerated, but

marked spasticity, such as we find in the spastic diplegias of childhood, is not common. In some instances there is an unusual sensitiveness to touch and to sound (hyperacusis), the child being startled by the slightest noise. Convulsions occur in some cases, but are not a frequent or integral symptom. All the functions of the body are in a low state of activity. The children are subject to frequent bronchial attacks, and often show gastro-intestinal disturbances. An examination of the fundus reveals the peculiar condition described by Tay. The "cherry-red spot in the region of the macula lutea" has become a familiar phenomenon to those who are on the lookout for this disease. There is a gradual increase of all the symptoms, the mental defect becomes more and more noticeable, the palsy more extreme, complete blindness follows, and the child lapses into a condition of marasmus in which it dies.

To sum up briefly, the chief symptoms are: (1) A mental impairment during the first months of life leading to absolute idiocy. (2) Paresis or paralysis of the greater part of the body, which may be either flaccid or spastic. (3) The reflexes may be normal, deficient, or increased. (4) A diminution of vision terminating in absolute blindness (the cherry-red spot in the region of the macula lutea and later a simple optic nerve atrophy). (5) Marasmus and a fatal termination, as a rule, before the age of two years. (6) The occurrence of the affection in several members of the same family.

In some, but not in all of the cases, nystagmus, strabismus, hyperacusis, and convulsions may be added to the above cardinal symptoms. A loss of the sense of hearing was noted in a few cases. Falkenheim referred to "explosive laughter," but the writer has not been able to corroborate this finding. If we allow for slight variations, it must be said that there are few diseases as uniform in their clinical manifestations as this. In a few cases, such as the one reported by Koller, the optic nerve changes preceded the characteristic changes in the macula lutea, and in one of Higier's cases the optic nerve atrophy was much more marked than the changes in the macula lutea.

**Diagnosis.**—The clinical symptoms are so distinct that a physician who is acquainted with the disease can hardly fail to make the diagnosis. The recognition must depend upon the concurrence of the various symptoms. Thus, there are innumerable forms of brain defect that do not belong to this category. There are also disturbances of vision, often syphilitic and of congenital origin, that have nothing to do with amaurotic family idiocy. But if a child that appears to have been well at birth loses its vision in the course of the first six months of life, if the child becomes listless, and if, on examination, the "cherry-red spot" can be found, there can be no doubt whatever about the disease. Some difficulty will occasionally be experienced in differentiating between amaurotic family idiocy and the congenital *diplegias* and *paraplegias*, but the preservation of considerable intelligence and the presence of normal vision ought to be sufficient to prevent mistakes. Children suffering from amaurotic family idiocy may have either flaccid or spastic limbs. On the whole, the flaccid limbs are fully as common. It is necessary to insist upon this since the writer has found that others seem to

consider spasticity a regular feature of the disease, which it surely is not. It is important in every suspected case to inquire into the character of the mother's labor at the time the child was born. Difficulties in labor have nothing to do with amaurotic family idiocy, but are commonly associated with congenital spastic palsies and with various forms of congenital idiocy and epilepsy. Convulsions may occur in amaurotic family idiocy, but do not constitute an integral symptom of the disease.

It is not always easy to distinguish *syphilitic* cerebral disease from amaurotic family idiocy, and if several members of the same family are afflicted, as they well might be in the case of an hereditary syphilitic disease, the difficulties become still greater.

Amaurotic family idiocy as described by the present writer is to be differentiated from the *juvenile form* to which Spielmeier<sup>1</sup> and Vogt<sup>2</sup> called special attention. This bears a close resemblance in many respects to the Tay-Sachs (infantile) type, but it occurs much later in life, often not appearing until the eighth, tenth, or twelfth year, is associated with blindness (without the changes in the macula), and leads to a fatal termination after a period of a few years. The writer has been struck by the resemblance between these two forms, and will not deny that they are akin to one another, without being identical. This kinship must also be upheld in view of Schaffer's investigations, which reveal a great resemblance between the histological findings of the juvenile and the infantile forms of amaurotic family idiocy. Such differences in histological structure as do occur might be due to the fact that the juvenile form represents a less violent process, attacking a brain farther advanced in development than the infantile brain of typical amaurotic family idiocy. While I would not deny that these two forms are somewhat related to one another, I cannot consider them identical.

**Treatment.**—Under this heading there is little to be said. As in the case of other family affections, we have no means of staying the disease. I have been asked by the parents of these "amaurotic" children, whether the birth of normal children can be expected. In several of the families that I have seen, normal children have grown up side by side with the unfortunate victims of the disease. In such families it has not been restricted to one sex or the other. From my own experience I can only advise that the dangers of blood relationship be seriously considered; that marriage between persons closely related be discouraged; and marriages between persons in whose families there are marked neurotic or psychical taints are also undesirable. If the disease has made its appearance in the child, purely symptomatic treatment is the only kind that can be adopted.

<sup>1</sup> *Neurol. Centralbl.*, 1906, xxv, 51; also *Monograph Gotha*, 1907.

<sup>2</sup> *Monatsschr. f. Psych. u. Neurol.*, 1906, xviii, 161.

## PART II.

# DISEASES OF THE LOCOMOTOR SYSTEM.

### CHAPTER XXIV.

#### MYOSITIS—MYOTONIA CONGENITA: THOMSEN'S DISEASE.

By WALTER R. STEINER, M.D.

#### MYOSITIS.

**Definition.**—Inflammation of the muscles. The voluntary muscles are, generally, alone involved, although the heart muscle may become affected, especially in the variety known as polymyositis hemorrhagica.

Myositis has long been recognized as occurring independently or in connection with certain diseases. Its classification presents considerable difficulty, for our knowledge of this subject is in a somewhat chaotic state. Dujardin-Beaumetz and others have endeavored to divide myositis into the acute and chronic varieties, which would greatly simplify matters. But unfortunately, as Lorenz pointed out, each variety is subdivided into acute, subacute, and chronic types, although the syphilitic and tuberculous forms have an especially chronic character. At present, Lorenz's division seems the most preferable, but we should remember that this classification may be only applicable for clinical convenience.

We shall limit ourselves to the consideration of the primary myositides. The examples of myositis found in pyemia, ulcerative endocarditis, actinomycosis, erysipelas, gonorrhœa, and other infectious diseases result from secondary processes. In tuberculosis, the so-called primary or hematogenous form is seen when the infection is carried to the muscles by the blood stream from a distant focus, and is not due to an extension from contiguous tissues. The secondary form is much the more frequent here, as Zeller,<sup>1</sup> after a careful search, could only collect thirteen typical cases of the former from the literature on this subject, and add two personal observations. Later, Kaiser<sup>2</sup> added six to this number and recently other investigators have increased the list to twenty-one cases.<sup>3</sup>

<sup>1</sup> *Beitr. z. klin. Chir.*, 1903, xxxix, 633.

<sup>2</sup> *Arch. f. klin. Chir.*, 1907, lxxvii, 1033.

<sup>3</sup> For a further consideration of this form see Dausel Ernst, *Ueber primäre Muskel-tuberkulose*, Berlin, 1912.

## PRIMARY SUPPURATIVE MYOSITIS.

**Definition.**—A single or multiple muscle inflammation, mostly acute, of bacterial origin, presenting clinically the picture of an acute infectious disease and generally ending in suppuration.

**Etiology.**—Suard was the first to conjecture that the real cause of this disease was of infectious origin, which Scriba<sup>1</sup> and Brunon<sup>2</sup> later proved by finding micro-organisms in the pus from the muscle abscesses. The results of the bacteriological labors of these observers were described by them in subsequent communications, but Scriba's work, appearing only in Japanese medical literature, has been pretty generally overlooked. The latter observed the *Staphylococcus pyogenes aureus*, while Brunon found streptococci, staphylococci, and an undifferentiated bacillus, the streptococci predominating. Brunon consequently thought that the disease was not due to a fixed variety of bacteria, but to any kind of pus-producing micro-organism. Other investigators, as Lorenz,<sup>3</sup> Honsell, Kojima, and Araki,<sup>4</sup> and Ito and Sinnaka,<sup>4</sup> have sought for bacteria in their cases, and have found, in every instance, the *Staphylococcus pyogenes aureus*. Miyake in a study of 33 cases obtained a pure culture of this organism in 27. Sato<sup>5</sup> twice obtained a mixed culture of staphylococci, streptococci, and an encapsulated diplococcus, resembling the pneumococcus; in another case, he cultivated staphylococci and streptococci.

Various portals of entry for the bacteria may be seen, such as small infected wounds, furuncles, or acne pustules, but, as Miyake states, they cannot be considered as the original foci of infection unless the bacteria they contain are similar to those found in the muscle abscesses. Occasionally the mucous membranes, apparently, permit the infectious organisms to gain entrance. From these different foci they are carried to the muscles by the blood stream, so the disease should be considered as a septicopyemic infection, with the abscess formation limited to the muscles, and the only reported blood culture confirms this theory. As muscles are very resistant to inflammation, the presence of a *locus minoris resistentiæ* is assumed, and various predisposing factors have been assigned.

**Age** appears to have no relation to its occurrence, although it was formerly thought that the disease was more frequent in children. In Miyake's series, 18 were under and 14 were over twenty-five years of age.

The *sex* bears no direct influence but it is seen more commonly in men, as in them the predisposing causes of trauma and overexertion are more frequently observed. In Miyake's series, 21 were men and 12 were women. In Ito and Sinnaka's list of 10 cases, 7 were men and 3 were women. It is more common in the laboring classes.

**Season.**—Some writers have thought the time of year, which has been variously given, had something to do with the onset. Fujiy claims it is most frequent when the laboring classes work in the fields and are subjected to the predisposing causes of cold and fatigue.

<sup>1</sup> Miyake, *Millh. a.d. Grenzgeb. d. Med. u. Chir.*, 1904, xiii, 153 (Bibliography).

<sup>2</sup> *Contribution a l'Etude de la Myosite infectieuse primitive*, Paris, 1887.

<sup>3</sup> *Die Muskelerkrankungen*, Vienna, 1898, 156.

<sup>4</sup> See Miyake, *op. cit.*

<sup>5</sup> *Deut. Zeitschr. f. Chir.*, 1903, lxi, 302.

**Pathology.**—Three varieties of suppurative myositis have been recognized: (1) With large solitary abscesses; (2) with disseminated abscesses, and (3) with diffuse purulent infiltration.

In primary suppurative myositis, most of the reported cases can be classed under the first heading. The skin and underlying connective tissue over the affected muscle or muscles are generally normal in appearance. The muscle itself is of a dark red to grayish red color, densely infiltrated with serum and very friable. The abscess, situated in the muscle, contains thick, greenish-yellow pus, mixed with blood and tissue particles. Its walls are lined with gray or yellowish necrotic fibres. In the more severe cases, where the abscess has invaded the whole muscle, only a few shreds of muscle tissue may be seen, hanging from the sarcolemma sheath (Chassaignac's case). Occasionally the muscle affected is described as pale, like the muscles of a fish, and exhibiting yellowish lines or streaks. It may also be bluish red in color, depending upon the degree and duration of the inflammation.

Microscopically the findings have varied somewhat. They are of a serous, seropurulent, and purulent character. Scriba declared that the interstitial tissue was not altered, but Miyake examined the muscle tissue in 14 of his 33 cases and observed the interstitial tissue markedly increased in the purulent cases and infiltrated with mononuclear and polymorphonuclear cells. Occasionally this tissue had completely replaced the muscle fibres.

The different investigators agree in the main on the muscle changes, but not upon their extent. Miyake found most of the fibres in his cases unaltered, while Scriba described the majority as swollen and showing a partial or entire loss of cross-striation. At times irregular cross-striations were noted. The sarcolemma sheath was much thickened and contained cells of varying sizes and shapes, while the perimysium was slightly infiltrated with small round cells. Hyaline, granular, and fatty degeneration was not noted by Miyake. Lorenz has described vacuolic degeneration. About the abscess walls there is a marked round-cell infiltration, and the muscle fibres are here almost completely destroyed. Farther out they are readily seen, but they are shrivelled and without cross-striation. The muscle nuclei are generally somewhat increased. By the Gram stain, bacteria are found in clumps in the vicinity of the abscess wall, but are not seen in localities where the inflammatory process is less pronounced. In advanced cases there is an extensive amount of granulation tissue and a new formation of muscle fibres.

**Symptoms.**—The onset is usually sudden, and most of those attacked have previously been in good health. A chill, followed by high fever, is generally the first symptom noted, but the patient may also complain of anorexia, general malaise, headache, profuse perspiration, and pains in the extremities. The pain is at first ill-defined but soon becomes localized in the muscles, which are very painful. On examination, one finds a tender, indurated swelling, which conforms to the shape of the muscle and is freely movable. Scriba describes the swelling as of a hard, board-like character. If it involves the long muscles, it is spindle-shaped in appearance, but if the broad muscles are attacked, it is hemispherical.

The muscle itself is contracted, and active and passive movements are much limited on account of pain. The skin over the affected muscle is hot but generally not reddened. There may be some œdema of the underlying tissues. At times an extensive erythema or ecchymosis is noted. The induration may be limited to only a portion of the muscle, or even its whole extent may be involved. Subsequently, within four to ten days, the swelling becomes softer and gives signs of evident fluctuation. In one case, however, this was delayed for thirty-one days. On incision, a muscle abscess is found and a thick, greenish-yellow pus, slightly mixed with blood, is evacuated. If the incision is long delayed or neglected, the pus may penetrate the muscle sheath and infiltrate the surrounding tissues, leading to the formation of multiple abscesses in different parts of the body, and finally result in death (Scriba). Rarely the stage of induration ends in resolution.

One or many muscles may be attacked. If multiple, it appears in two forms: (1) The so-called typical form, which is multiple from the onset; (2) the atypical form, which begins as an isolated focus of infection and later becomes multiple (Miyake).

Recurrences may take place. Two cases have been reported in which this happened after an interval of three and four months. The similarity between this disease and osteomyelitis, both clinically and bacteriologically, has been especially considered by Walther and Miyake. Brunon divides the cases into three types, viz.: (1) Malignant, resulting in death in a few days; (2) acute, in which recovery may ensue; and (3) subacute, a very light form. Some of his cases belong to other diseases, and a consideration of the typical instances of this affection, thus far reported, leads us to place them under two headings, the acute and subacute.

**Diagnosis.**—If the disease is seen from its outset, this is generally relatively simple, provided the following two points be considered: (1) The dense infiltration and swelling of the muscle; (2) the painful condition of the affected muscle, which is kept firmly contracted, and causes the extremity, or part of the body implicated, to assume various positions. If, for instance, the biceps brachii is attacked the arm will be flexed, or, if the rectus abdominis muscle be involved the body will be inclined forward. In each case the insertions of the muscles are more closely approximated, to lessen the pain which tension would increase. Occasionally osteomyelitis is simulated, especially in those cases in which the muscle sheaths of the implicated muscles have ruptured. In the latter, the disease rarely involves the surrounding tissues, but is seen extending to other parts of the body by metastatic foci. In doubtful cases, a wide surgical incision, combined with an inspection and palpation of the abscess cavity will definitely differentiate the two conditions. Metastatic abscesses in pyemia will at times need to be distinguished from this malady. In the former there is usually no dense infiltration of the muscle or muscles, or, if present, it is extremely slight. The local disturbances are absent or so trivial that the patient does not detect them.

**Prognosis.**—All uncomplicated cases have a favorable prognosis if seen early enough for the requisite surgical treatment. In Miyake's series of 33 cases there was only one death, and this was from a complica-



tion. After the pus is emptied from the muscle abscesses, the acute inflammatory symptoms subside very promptly, and complete healing takes place in the affected muscles, after an interval of from one and a half to three months. Rarely, the loss of muscle substance is replaced by scar tissue, which may cause severe contractures of the muscles. In cases in which early surgical treatment has been neglected, if the pus penetrates the muscle sheath and forms metastatic foci in other parts of the body, the prognosis becomes very grave. Muscle atrophy may follow the healing of the abscesses, but there is generally no subsequent functional disturbance.

The complications to be dreaded are pyemia and pneumonia. Sato, Kurosawa, and Suzuki have reported cases in which a metastatic lung abscess was the cause of death. In Walther's case death ensued in the ninth week from an intercurrent endomyocarditis.

**Treatment.**—By the use of cold packs and other antiphlogistic means, the muscle induration may not undergo suppuration, and absorption may result. If fluctuation does occur, an early broad incision should be made and the pus thoroughly evacuated. It should be remembered that a previous negative exploratory puncture does not exclude the presence of pus, which may have been too thick to flow through the needle. Fujiy and Honsell oppose surgical procedures unless pus is surely present, but in case of doubt it is better to err on the side of caution and open the indurated areas widely. If, after the myositis has subsided, contractures ensue, massage and various orthopedic measures should be employed.

### NON-SUPPURATIVE MYOSITIS.

**Dermatomyositis.**—**Definition.**—An acute, subacute, or chronic disease, of unknown origin, characterized generally by a gradual onset, with vague and indefinite prodromes, followed by oedema, dermatitis, and a multiple muscle inflammation.

**Etiology.**—We are much in the dark as to the cause, although recent findings point more strongly to its infectious origin. Hepp and others have previously emphasized this view, and the presence of a splenic tumor, fever, and angina have seemed to support it. In most of the recent cases bacteria have been found, organisms similar to staphylococci being described but these cases appear generally atypical. Senator advanced a toxic theory, which has, at present, no supporters.

**Distribution.**—Cases have been reported in the United States and in a number of European countries.

**Race.**—The Anglo-Germanic has furnished most of the instances, followed by the Latin and Scandinavian races. The case reported by the writer is the only one yet observed in the negro.<sup>1</sup>

**Season.**—The time of year seems to have no connection with the disease. In the writer's collected series, 10 were attacked in the winter, 5 in the spring, 8 in the summer, and 5 in the autumn.

<sup>1</sup> *Jour. Exper. Med.*, 1903, vi, 407 (Bibliography).

*Sex.*—The affection has been observed, according to this series, in 17 males and 11 females.

*Pathology.*—Of the 17 fatal cases in this series, autopsies have been performed in all but 7, and in 4 of these the excision of a piece of muscle was allowed. We have, consequently, a fair amount of data on which to base conclusions. With the exception of an enlarged and soft spleen, the pathological findings are limited to the muscles. Any or all may be attacked. It was early stated that the muscles of the eye, tongue, heart, and diaphragm were exempt, but later investigation has shown they may become implicated. Lorenz<sup>1</sup> declares the implication of the heart has a deeper significance than was at first imagined. The cases presenting this complication show a special tendency to relapses, although mild and abortive at first.

The skin covering the muscles is firm and hard and generally does not pit on pressure, although œdema is usually present, but may be slight. On section the subcutaneous tissues present a firm, tense œdema, and are, as a rule, infiltrated with a yellowish, serous fluid. The muscles exhibit extensive changes; they are swollen, pale red or pale yellow in color, or may reveal, occasionally, yellowish-gray or diffuse reddish streaks. Hepp considers the muscles to resemble those of a dog. They are often strongly infiltrated with serum and quite moist. In consistency they vary, being hard and firm, or soft and boggy. They may be quite friable, as in Hepp's case, in which the left rectus muscle was found ruptured at autopsy. They are without lustre and of a dull, opaque appearance. Hemorrhages may occasionally be seen in them. Microscopically the changes are those of a parenchymatous and interstitial inflammation, and may vary in extent, being either focal or diffuse. Again, the different muscle changes may be seen, occurring in one and the same fibre. These are frequently separated from one another by the existing œdema or by mononuclear and polymorphonuclear leukocytes. Small hemorrhages may also be seen between them. The fibres are found in all stages of degeneration; they may be normal in size, œdematous or atrophied, coarsely or finely granular, hyaline or waxy, occasionally fatty. The striæ are normal, indistinct or invisible. Longitudinal or cross-cleavage of the fibres has been found, and vacuoles have been described in four instances. In many cases there is an increase in the number of muscle nuclei. Typical interstitial foci of small round cells are found in the perivascular connective tissue and, to a lesser extent, between the muscles. In the subacute and chronic cases the increase in connective tissue may be marked in both the perimysium externum and internum, and solitary muscle fibres in the process of degeneration may be surrounded on all sides by connective tissue. In five cases the bloodvessels were somewhat dilated and filled with blood. In seven of the autopsies, bronchopneumonia was found as the terminal infection.

*Symptoms.*—The disease generally attacks persons in the prime of life and in the best of health. Fuckel's case, however, was noted in a girl after an attack of measles, and both of Wagner's cases had pulmonary

<sup>1</sup> *Berl. klin. Woch.*, 1906, xliii, 727, and *Deut. med. Woch.*, 1906, xxxii, 777.

tuberculosis, the second patient having tuberculous ulcers of the intestines. Senator's second case was a diabetic. The onset is almost always gradual, but may be sudden, with the prodromal symptoms of malaise, weakness, anorexia, pains in the extremities, or headache. These symptoms may be of several days' to three weeks' duration, or even longer, as in one case. Occasionally they are absent.

*Pain.*—Vague pains are next complained of, as well as a stiffness or rigidity in the extremities and back. These pains quickly take on a more definite character, become more or less circumscribed, and are localized in the muscles. Different muscle groups are then successively attacked, and eventually the whole skeleton musculature may be implicated. Later in the disease the pains become more severe, and are not only spontaneous in origin, but also are caused by active and passive movements. They are described as drawing, tearing, or boring in nature. In the most severe cases the patients lie utterly helpless in bed, as if completely paralyzed. The muscles involved are generally very painful on palpation, although the cedema does not cause them to be well defined. Muscle contractures have been observed in a few cases.

*Fever* is soon noted. It is usually of moderate intensity and intermittent or remittent in type. It rarely exceeds  $104^{\circ}$ , but just before death may rise several degrees above this height.

*Edema.*—This appears with the fever and may implicate the whole body and extremities, the latter presenting at times a most ungainly appearance. It is generally first seen on the face, especially above the eyelids, and may cause the countenance to assume an immobile aspect, likened by Oppenheim to alabaster. The swelling is, as a rule, always noted in the extremities, being apt to involve the proximal parts, *e. g.*, the shoulder, upper arm, elbow, thigh, or inguinal region. It is not a symmetrical cedema, as the flexor or extensor surfaces are generally alone involved. It also varies greatly in character, as at one time it may pit on pressure, while at another a dense, hard infiltration of the skin is observed, the latter being the more usual. After the skin inflammation is noticed, it becomes more intense, and may remain localized over the affected muscles or spread to surrounding parts. Usually its duration is limited to the acute stage of the disease. The wrist- and ankle-joints are generally spared.

*Dermatitis.*—This is an early symptom, and varies greatly in character, being in different cases an erythema, a pseudo-erysipelas, an urticaria, a roseola, an eczema, or an inflammation resembling erythema nodosum. It may spread continuously or remain limited to the parts where it was first observed. At times it occurs only later in the disease. Its location is apt to be over the diseased muscles. It may disappear without leaving a trace behind, or a pigmentation may indicate its former presence. In two instances, a dermatitis of a different type followed the disappearance of the first eruption.

Profuse perspiration and an enlarged spleen usually accompany the other signs and symptoms.

*Nervous System.*—No disturbances of sensation are, as a rule, met with, and the nerves are not tender on palpation. Hepp refers to peculiar

cramp-like pains in the later stages of the disease, of variable duration, and Wagner, Lewy, Lorenz, and Leube have noticed paresthesia during the course. This, with the cramp-like pains, was especially marked in the writer's case, in which the patient complained of a sensation as if something was crawling down from the elbows of each arm to the fingers; it was particularly noticeable at night. The knee-jerks and the electrical reactions are usually either normal or diminished. In the mild cases the knee-jerks may be slightly increased, and, in the most severe instances, they may be totally abolished. Oppenheim states that there is, generally, a quantitative decrease in the electrical excitability of the muscles, and Lewy got a partial reaction of degeneration in his case.

Stomatitis and angina are at times seen, either early or late in the disease. In Lewy's case and in four of Oppenheim's, ulceration of the mucous membrane was observed. On this account the latter investigator has coined the word *dermato-mucoso-myositis*, to name this disease.

The extension of the affection to the muscles of respiration and deglutition was formerly considered the rule, and Lowenfeld names this as one of the three cardinal symptoms. It well accounts for the number of fatal cases reported, due to suffocation or bronchopneumonia. The urine is usually normal, but may contain albumin, and hyaline and granular casts may be found in the urinary sediment. The course may be characterized by improvement and relapses in the subacute and chronic cases, in which atrophy of the muscles may be observed. The acute cases last from one or two weeks to two months; the subacute, two to eight months, and the chronic forms from a year and a half to two years (Martini, Lorenz).

**Diagnosis.**—As a rule, no difficulty will be experienced in typical cases. Diseases presenting somewhat similar symptoms are: (1) Trichinosis, (2) neuromyositis, (3) primary suppurative myositis, and (4) syphilitic myositis. In the first, the initial gastro-intestinal disturbances and the discovery of trichinae in the stools and excised muscles; in the second, the chain of nervous phenomena and the absence of a dermatitis; in the third, the presence of a focus of infection with the bacteriological report on the muscle examination; and in the fourth, the history of the patient and the objective findings, will generally sufficiently differentiate the affection. It is also well to bear in mind that one of Oppenheim's cases turned out, four years later, to be a typical example of scleroderma, while another in his series was so diagnosed by the physician who first attended him.

**Prognosis.**—As all the muscles of the body may be implicated, including those of respiration and deglutition, death may result from suffocation or bronchopneumonia. The early cases of this disease were almost invariably fatal, but some of those reported later have resulted in recovery. Of the 28 cases collected by the writer, 17 terminated fatally, so the prognosis is always grave. The outcome was fatal for the two patients in both extremes of life.

**Treatment.**—This should be chiefly directed to the relief of pain and to the keeping up of the patient's nutrition. Various analgesics have been tried for the former object, and among them we may mention aspirin

and the salicylates, but no particular drug has as yet been found to offer any especial advantage. In severe cases, morphine has sometimes been necessary, to quiet the pain. Oppenheim employs hot air, the use of hot drinks and aspirin, while every second day he endeavors to cause the patient to perspire profusely. At the onset, or later, he uses thermomassage, and subsequently massage, exercises, and electrotherapy.

**Polymyositis Hemorrhagica.**—**Definition.**—An acute, subacute, or chronic disease, of unknown origin, strongly resembling dermatomyositis, but differing from it chiefly in the presence of a greater or less amount of interstitial hemorrhage between the muscles and the occurrence of circulatory symptoms, caused by the implication of the cardiac muscle.

**Etiology.**—This is quite obscure, but staphylococci were found in two instances. In two cases, cold and fatigue seemed to bear some relation to the onset; in another case, angina was the first symptom, and the disease was thought to be a general infection following it. In still another, there was some preceding inflammation of the tissues of the neck.

**Pathology.**—The findings vary from simple hemorrhages between unchanged muscle fibres to extensive muscle degenerations and new connective-tissue formation. Lorenz has divided the changes into the acute and chronic forms. The first is characterized by intramuscular hemorrhages, which cause destruction of the fibres, as well as the many muscle degenerations described under dermatomyositis. The second form is distinguished by the presence of connective tissue containing blood pigment and markedly atrophic fibres. For a fuller discussion of the pathology, one is referred to the article on Dermatomyositis.

**Symptoms.**—These described under dermatomyositis may be well used here, although in polymyositis hemorrhagica, the usual absence of prodromal symptoms, a more sudden onset, and the occurrence of less fever are seen. *Pain* is, generally, the first symptom noted; it is definitely located in a small circumscribed area in the muscles, where, later, a nodular tumor, tender on palpation, develops. The extremities are usually first attacked. The condition is accompanied by more or less oedema, which varies greatly in its characteristics, for it may be as extensive as in dermatomyositis, but frequently it is more circumscribed and of softer consistency. Different muscle groups are gradually attacked, and the whole skeleton musculature may be finally involved. The skin may show a hemorrhagic or a measly eruption. The former is revealed by the presence of violaceous and purpuric spots, which, when fading, leave behind a yellowish-green discoloration. Pigmentation may eventually be noted. Prinzing's<sup>1</sup> case exhibited well-marked muscle atrophy. Circulatory symptoms have been described in every case save one, due to the implication of the cardiac muscle. They are cardiac palpitation, tachycardia, arrhythmia, and, more rarely, murmurs; all of which account for the cardiac weakness and collapse so frequently observed. An enlarged spleen is an inconstant symptom as well as hemorrhage, apart from that into the muscles and skin. Bleeding from the intestines has been described, and attacks of epistaxis, hematuria, and profuse menstrual

<sup>1</sup> *Münch. med. Woch.*, 1890, xxxvii, 846.

flow. Nephritis is a frequent complication. The disease is generally of several months' duration. Fenoglio's<sup>1</sup> patient lived on for a year and a half, finally dying of pulmonary tuberculosis.

**Diagnosis.**—The sudden onset, the moderate fever, the character of the skin eruption, the cardiac involvement, and the muscle findings will readily cause it to be distinguished from dermatomyositis or other seemingly similar affections.

**Prognosis.**—Four out of the ten cases collected by Thayer recovered,<sup>2</sup> so that the prognosis is about as grave as in dermatomyositis.

**Treatment.**—See Dermatomyositis.

**Neuromyositis.**—This term has been applied in about ten instances to cases which present the signs and symptoms of a myositis associated with those of neuritis. There seems to be some uncertainty as to whether the disease exists *per se* or whether the co-existence of two diseases is here observed. It is usually seen in chronic alcoholics and accompanied by ataxia.

### MYOSITIS WITH SPECIAL TERMINAL LESIONS.

**Primary Myositis Fibrosa.**—**Definition.**—A single or multiple inflammation of the muscles, mostly subacute or chronic, which generally begins in the lower extremities, and presents but slight constitutional symptoms. Eventually the muscle tissue concerned is largely or wholly replaced by connective tissue, and quite pronounced muscle atrophy may be then observed.

**Etiology.**—There has been considerable discussion whether this disease in its primary form is a definite entity. Some have claimed that it is only seen secondary to infectious or rheumatic conditions, but there seem to be good grounds for considering its independent existence as established. Cultures from the tissues and search for micro-organisms in sections have been uniformly negative.

**Pathology.**—The changes in the muscles are seen in two forms. In the first the implicated muscles are swollen and present to view a tumor-like mass. On palpation, they are firm and hard. The skin and underlying tissues may be involved also, and then a dense oedema appears which does not pit on pressure. A brown pigmentation of the skin has been at times observed. In the other form, the muscle is changed into a dense, tendinous band. On section the affected muscles grate under the knife and reveal a hard white surface in the most involved areas, but in the less affected portions show reddish-yellow spots, which represent the remains of some of the muscle fibres. Microscopically there is a great increase in the interstitial tissue, which, in places, entirely replaces the muscle fibres. In other portions the fibres are atrophied and show a granular or fatty degeneration. Their cross-striations have disappeared, but their longitudinal striations are more evident. A relative increase of tendinous tissue is at times seen.

<sup>1</sup> *Rivista clin. Arch. ital. di klin. med.*, 1890, p. 497.

<sup>2</sup> *Boston Med. and Surg. Jour.*, 1902, cxlvii, 312.

**Symptoms.**—The onset is usually slow and with slight symptoms. Indeed, the disease may take from several months to ten years to develop. Sharp pain is first complained of in the affected muscles, those of the lower extremity being usually the first implicated. Only one muscle or a single group of muscles are generally first attacked, although the beginning of the malady may be in several muscles. The pain in the concerned muscles soon forces the patient to go to bed, and, subsequently, a rigidity of the affected limbs results and contractures ensue, the flexor muscles being especially prone to involvement. There is no fever, and the pain usually present is only of slight duration. The general condition is but little altered. Finally the affection may implicate most of the voluntary muscles of the body. The electrical excitability of the muscles is reduced, or may, in extreme cases, be wholly wanting. Disturbances of sensation have only rarely been reported.

**Diagnosis.**—As Lorenz has well said, a positive diagnosis is impossible without histological examination. The slight or total absence of pain, when the affected muscle is palpated, is said to be characteristic as well as the gradual hardening of the muscle implicated and the cessation of the spontaneous pains, which are first well marked.

**Prognosis.**—The duration is long, but eventually a decided improvement is usually noted. However, it is ever well to remember that the affection may remain stationary.

**Treatment.**—Drugs are of no service, but massage and electricity are of benefit in ameliorating the symptoms and hastening the subsidence of the disease. By their use recovery has been noted.

**Progressive Myositis Ossificans.**—**Definition.**—A progressive inflammatory affection of the locomotor system, of unknown origin, characterized by the gradual formation of bony masses in the fascia, muscles, aponeuroses, tendons, ligaments, and bones, with resulting ankylosis of most of the articulations.

**Etiology.**—In spite of careful study the cause is as yet unknown. It is frequently described as of congenital origin, in the sense that it depends on some aberration of growth, congenitally acquired. Fanning<sup>1</sup> reported a hereditary case but it is unique in the literature. There are, at present, two main theories. The first one considers the process of bone formation to be of an inflammatory nature, while the second view regards the bony growth as representing a tumor formation. Goto,<sup>2</sup> accepting neither theory, considers the initial process to begin with a connective tissue hyperplasia which leads frequently to the final stage of bone formation, through one of fibrous induration.

**Age.**—The disease is generally first noted in young persons, as Lorenz's records show, for out of 45 cases in which the age at onset is given, 38 were affected before the fifteenth year; 11 of these showed the disease during the first year of life, 16 between the first and fifth years, and 11 between the ages of five and fifteen. Occasionally the patient does not fall a victim until later in life. Roger's patient was thirty-five years old when stricken. It appears to begin earlier in women than in men.

<sup>1</sup> *Lancet*, 1901, ii, 849.  
VOL. V—56

<sup>2</sup> *Arch. f. klin. Chir.*, 1912-13, c, 730.

*Sex.*—Males are more frequently attacked than females. Gerber gives the ratio as 4 to 1, while Rolleston says it is 5 to 1. In Weill and Nissim's series of 50 cases, 38 were men and 12 were women. To account for this, Pincus lays particular stress on obstetrical traumatism, and says the more frequent injuries of boys at birth explains the preponderance of the male cases.

*Race.*—The Anglo-Germanic race has furnished most of the instances. According to Weill and Nissim's series, 20 cases came from Germany and 19 from England. The Latin and Scandinavian races contribute a few instances, and Russian, Swedish, and French cases have been reported.

*Pathology.*—In 1869 Münchmeyer described the process of bone formation in the muscles. He recognized three periods or stages which have been followed by other writers: (1) Stage of embryonic infiltration; (2) stage of connective-tissue induration; and (3) stage of ossification.

In the first stage the skin and subcutaneous tissue may appear normal, but the underlying affected muscle is much swollen and œdematous. Microscopically an extensive infiltration of the intramuscular and intermuscular connective tissue is seen. This infiltration consists in the formation and proliferation of an embryonic connective tissue, and is especially marked about the blood vessels. In the second stage the embryonic connective tissue becomes organized and forms adult connective tissue, which, at first, proliferates more and more, but finally retracts and becomes a hard, fibrous swelling. On section it has the appearance of a fibroma, presenting a hard, white mass, in which the remaining fibres resemble red striations. These fibres are more numerous at the periphery of the tumor where, microscopically, many of them appear normal. On passing inward toward the centre of the hard, white mass, the fibres, when present, seem atrophied or swollen, and frequently their cross-striations are lost. Fatty, waxy, and granular degenerations have been described, and many fibres are completely destroyed. Their sarcolemma nuclei may be markedly increased and some may resemble giant cells. The increased intermuscular connective tissue is very rich in cells from the fusiform up to the large polymorphous type, which contains numerous karyokinetic figures. Other cells are also seen which resemble cartilage cells, without capsules, and later produce hyaline cartilage. Toward the middle of these fibrous swellings, small angular spaces are observed, in which formative cells lie, which later represent osteoblasts and bone corpuscles. The spaces are the osteoid trabeculæ. When these become calcified, the parts enclosed by the trabeculæ become bone-marrow, and true compact and spongy bone is formed, when ossification, seen in the third stage ensues. New bone is also formed in the fasciæ, ligaments, tendons, and preëxisting bones, in a manner similar to that just outlined. It is always of the fibrous, cartilaginous, or periosteal type, and histologically and chemically resembles true bone. By the fusion of several bony foci a larger mass is produced, and eventually a bony tumor, situated originally in a muscle, may extend beyond the confines of the affected muscle and give no indication of its point of origin.



Friedberg, and Salmann<sup>1</sup> are opposed to this sequence of histological changes. They regard the disease as a primary parenchymatous muscle inflammation, with a secondary participation of the intramuscular and intermuscular connective tissue. It is proper to state that but one muscle was affected in the latter's case, and the sections were differently interpreted by Lexer when he had the opportunity to examine them, for he considered the connective tissue changes to be primary. Goto considers the process to begin in the fascia and would name the affection hyperplasia fascialis ossificans progressiva.

**Symptoms.**—The disease may exist for years before it is recognized. Many cases, indeed, have been diagnosed as "rheumatism," and their true nature only revealed at the period of bone formation. At times the onset may be so insidious that inflammatory appearances in the affected muscle, as well as pain, may be completely lacking. In typical cases the signs of a myositis are present; without any known exciting cause or after trauma, which may be very insignificant, a swelling is found in the muscle involved. The swelling presents a firm, doughy consistency. Pain may be present, confined to the muscle attacked or radiating in character. There is œdema of the overlying tissues, with some redness of the skin. The temperature is usually slightly elevated. In the course of a few days, or longer, depending upon the intensity of the process and the extent of the lesion, these symptoms vanish, but the muscle does not return to its normal state. It remains indurated, and finally a hard, tumor-like mass results. The disease may then become quite stationary for a variable period of time. Eventually, many of these masses form bony deposits, but some may remain as fibrous tumors, while others disappear, causing the muscles to undergo fibrous degeneration. In such muscles, Münchmeyer and others have described the occasional presence of fibrillary contractions. More rarely, the muscle may be entirely restored to its former state by the disappearance of the tumor. The process of bone formation occupies no fixed time, generally it is from two to eight months (the latter duration, being seen in Koht's case). The bony change may be observed in the whole muscle or only in its fleshy portions, or, exceptionally, the tendons alone may be concerned. According to Münchmeyer, those muscles are spared whose two extremities are not inserted in the skeleton, but subsequent investigations have not borne out this statement. The condition is usually painless, although, occasionally, spontaneous pains are complained of, especially at night.

The muscles of the back and neck are generally the first involved. They were the first implicated in 21 out of 38 cases, while the muscles of the upper and lower extremities and of the face were the first attacked in the other instances (Lorenz). The ossification of the cervical ligaments and the muscles of the neck causes the head to be bent forward and the neck to be fixed. In extreme cases the flexion is so marked that the chin touches the sternum. The ossification of the ligaments, fasciæ, and muscles of the back varies very greatly. The bony deposits, if involving only a muscle, may conform to its shape and feel, on palpation, like

<sup>1</sup> *Ueber die Myositis ossificans progressiva*, Berlin, 1893.

freely movable, irregular plates. Very often the ossification has extended farther and the muscles are firmly attached to the skeleton. Generally, bony foci may be recognized in many muscles of the back, with no regular arrangement, like the ramifications of coral (Copping) or a geographical map of a mountainous region (Brennsohn). Minkewitsch describes the bony tumors of the back as resembling a stag's antlers. The vertebral column eventually becomes fixed, by the formation of bridges of bone, and scoliosis may be observed. A kyphosis, either cervical or lumbar, is much more rarely seen. The scapulæ early become attached to each other and to the thorax, while occasionally they are connected with the ileum. The movements of both arms consequently are much limited. If the pectorals and latissimus dorsi are also involved, the arms are rigidly fixed on the thorax, which in some cases is described as a bony coat of mail.

Subsequently, the disease progresses, more muscles become implicated, and the corresponding joints fixed. The above outlined train of symptoms then ensues, except that fever may not be present. The upper extremities are usually earlier attacked than the lower, and a cicatricial shortening precedes the ossification, so that the limbs are held in a contracted position. The arms are generally fixed at the shoulder and, by the ossification of the biceps and anterior brachial, flexed at the elbow. The fixation of the ilio-femoral articulations, as well as the ossification of the gluteal muscles, cause the hips also to be slightly flexed at the pelvis. This posture, with the forward projection of the head, displaces the patient's centre of gravity and gives them a precipitous gait, their knees being generally fixed in extension or slightly flexed. The forearms, hands, legs, and feet are only rarely involved. If the lower intercostal or abdominal muscles are implicated, the respiration may be upper costal in type.

Finally, the temporals, masseters, and pterygoids are attacked, and the patient is unable to move his lower jaw, which may be somewhat atrophied at this period. To overcome the difficulty in feeding thus induced, teeth and a portion of the alveolar process of the lower jaw have been removed. The mimicry facial muscles, the muscles of the eye, tongue, diaphragm, heart, perineum, genital apparatus, and sphincters are unaffected. The electrical excitability of the muscles varies. With the galvanic the anodal closing contraction appeared before the cathodal closing contraction in two cases. The response to the faradic has generally been described as normal or decreased. Pinter got a reaction of degeneration. In women who contract the disease, the menses generally cease, or, if they have not yet appeared, they are frequently long delayed.

The presence of exostoses has been frequently described. They are usually seen on the internal surface of the arm, the anterior aspect of the tibia, the upper part of the fibula, the ribs, or where the long bones approach the skin. The frontal bones and the phalanges of the fingers are also other sites where they have been observed. They may subsequently disappear. In about 75 per cent. of the cases reported, a peculiar deformity of the great toes and thumbs has been noted, and occasionally

the little fingers also have been involved. This was first described by Gerber, but Helferich<sup>1</sup> directed more particular attention to it. The toes and thumbs are much shorter than normal, due to the dwarfing of the metatarsal and metacarpal bones, and subsequent ankylosis of the interphalangeal joints. This gave the impression, before the introduction of x-rays, that one phalanx was absent. The great toes are, generally, directed outward and frequently lie under the second toes, giving rise to the deformity of hallux valgus. The condition of the toes and thumbs is known as microdactylia. It is interesting to note that it was observed in the father of Sympton's patient, although he was not subject to this disease. In Michelsohn's patient the deformity was seen in the toes at birth, but on the thumbs it was not detected until the seventh year. Other malformations, such as lack of some muscles, atrophy of the testicles and scrotum, have been noted in individual instances.

**Diagnosis.**—No difficulty should be experienced in recognizing this condition in advanced cases. The age of the patient, the first localizations of the bony deposits, the ever progressing character of the disease, and the presence of exostoses and microdactylia are important factors in establishing a diagnosis. Multiple exostoses may sometimes be mistaken for it, but the location of these tumors at the diaphyses and epiphyses of the extremities, the arrest of their development past the age of twenty, their bony character from the onset, and the fact that the muscles are never involved, except secondarily, should suffice for a proper differentiation. The joint involvements seen in locomotor ataxia should never be confused with this malady, although some muscles near the affected joints occasionally undergo extensive ossification. In the disease we are discussing, the joints are never implicated. Again, spondylitis deformans and the muscle dystrophies bear some relation to progressive myositis ossificans, but their proper differentiation should present no difficulty.

**Prognosis.**—The disease is essentially of chronic character, and relapses alternate with pauses in its progress. At times there appears to be a cessation of the symptoms for six, ten, or even twenty-three years (as in Koht's case); but the malady is ever a progressive one, and the slightest cause may induce the formation of bony masses, such as the mere palpation of a muscle. In one patient a relapse occurred in April for five successive years without any known cause. Eventually the patients become practically helpless and bedridden. Bedsores and abscesses about the bony deposits may result, and death may ensue from pyemia or some intercurrent infection, such as tuberculosis, to which the patients seem susceptible, or pneumonia. Suffocation may intervene, due to the limitation of the costal breathing and the onset of œdema of the glottis.

**Treatment.**—Medical treatment avails nothing. Hawkins, Gibney and others had recourse to surgical measures, and some of the bony tumors were removed. The result has been most unsatisfactory, as there has been a recurrence at the site of removal in almost every instance. Still, in necessary cases, as when the lower jaw is fixed, operative measures

<sup>1</sup> *Aerztl. Intelligenzbl.*, 1879, xxvi, 845. For an excellent consideration of this deformity see Jungling, *Beitr. z. klin. Chir.*, 1912, lxxviii, 306. He has collected 29 cases and adds one from personal observation.

are justifiable. The most important point is prophylaxis. By exercising precautions against trauma and cold, the affection can frequently be prevented from spreading. Occasionally, even these measures will not check the progress.

### MYOTONIA CONGENITA: THOMSEN'S DISEASE.

**Definition.**—An affection characterized by tonic muscular cramps at the beginning of voluntary movements.

The disease was named after a Danish physician, J. Thomsen, of Kappeln, in Silesia, who was a sufferer from it, and in whose family it had existed for five generations.<sup>1</sup>

**Etiology.**—Heredity is an important factor, for the disease may be directly transferred from the patient to the offspring, or merely the disposition to the malady may be transmitted. It may also be seen in the collateral branches of the family, or occur in several members of the same family, without direct heredity. In 1890 Hershill<sup>2</sup> stated that there were fifteen families reported in which the disease thus existed. Bernhardt thinks that the marriage of blood relations may be of some etiological moment, as two members of the same family, whose parents and grandparents were cousins, came to him with the disease.

Occasionally, without heredity or family taint, the disease comes on in childhood, and has been found associated with a fright, a fall, or a dog-bite. These factors are probably not the cause, however, and we may conceive of the malady as being overlooked, on account of the mildness of the symptoms, until an accident first attracts particular attention to the child. Apart from these, other cases exist in which the symptoms have not appeared until later in life. In some instances, as in Thomsen's family, there is a marked neurotic family history, but Hale White<sup>3</sup> does not consider this to have much bearing on the etiology and adds that the patient himself is rarely of neurotic habit.

**Age.**—The disease generally comes under observation when the patients are about twenty years of age, but cases are not rare at puberty. If the symptoms develop in young children, they usually pass unnoted until puberty is reached. Then, for the first time, an awkwardness in different movements and an inability to take part in games is observed. Sometimes they find they cannot get up quickly when called upon to recite, or a soldier's career may first reveal the condition by the clumsiness and awkwardness with which the different movements are executed. When a diagnosis has been made in infancy, as in Friis' case, a difficulty in sucking, an immobility of the countenance after weeping, an awkwardness in bodily movements, or a panting respiration may be observed.

**Sex.**—Males are much more frequently attacked; in Niedendarp's list of 58 cases, 52 were men, while only 6 were women. In Hans Koch's list of 102 instances, this inequality was likewise very apparent, as 91 were men, but only 11 women. Of the 91 men, 11 had been soldiers.

<sup>1</sup> *Arch. f. Psychiat.*, 1875, vi, 706.

<sup>2</sup> *British Med. Jour.*, 1890, i, 242 (Bibliography).

<sup>3</sup> *Albutt's System of Medicine*, 1899, vi, 467.

*Race.*—Cases have been reported in Germany, France, Italy, Russia, Sweden, England, and in the United States. It appears to be more common in Germany and Scandinavia than elsewhere.

*Pathology.*—The material for the study of this disease has come mostly from muscles excised during life, as but one autopsy has been performed (Dejerine and Sotta<sup>1</sup>). The patient succumbed to an attack of acute nephritis with uremic symptoms. During the year prior to his death he was attacked by a chronic, exfoliative dermatitis, but otherwise had been in good health. The central and peripheral nervous systems are said to have been normal, but the cerebrum and cerebellum were not microscopically examined. There was a marked serous infiltration of the entire body, hydro-thorax, hydro-pericardium, and œdema of the lungs, besides the special muscle changes. In the earlier cases, before Erb's monograph was published, the examinations of the muscles were negative, although they were investigated in five instances. In 1886 Erb's work appeared, in which he detailed the complete histories of two patients, with protocols on the examinations of the portions of muscles taken from each of them. He also gave a report on some muscle sections from a man who had died soon after an operation. These sections showed pictures so similar to those seen in his two patients that he investigated the man's past history, and found him to have had Thomsen's disease.

In Erb's cases the marked *hypertrophy* of the muscles affected was very noticeable while the skin over them seemed normal. On palpation the muscles possessed a certain elasticity and appeared to be in a somewhat contracted state. Some of them even revealed a board-like hardness. When called into action they felt much firmer than a normal muscle would feel under the same circumstances. Their color on section offered nothing distinctive. Erb claimed he could distinguish with the naked eye the solitary muscle fibres. Microscopically, the stained specimens exhibited changes most marked in the muscle fibres, although the connective tissue was also somewhat implicated. The fibres, individually, showed considerable hypertrophy, some of them being twice and even four times the diameter of normal fibres. In fact, 70 to 75 per cent. of all the muscles measured from 60 to 140 $\mu$ , in comparison with the normal measurements of from 20 to 65 $\mu$ . The largest fibre had a diameter of 195 $\mu$ . On cross-section the bundles of fibrillæ did not retain their normal polygonal form, but were much rounder than usual, with blunt edges, some being even almost circular in shape. The fibres were also more homogeneous than normal, as there was an evident loss of cross-striation. Longitudinal sections showed a lack of the normal parallelism; they were curved and bent in all directions and provided with swollen, irregular borders. The cross-striations appeared finer and more delicate than usual, while occasionally they were wanting and pronounced longitudinal striations were noted, or the fibres presented a very homogeneous aspect. In one of his cases, vacuoles were present but they were not very frequent. They were of varying sizes, sharply marked off from the muscle fibre containing them, and generally somewhat eccentrically

<sup>1</sup> *Rev. de Méd.*, 1895, xv, 241.

placed. In shape, they were inclined to be ovoid. They contained a finely granular, homogeneous mass as their contents, or a mass appearing as a membrane or coagulum provided with smaller vacuoles. Apart from these abnormalities, the increase in the number of nuclei was even more striking. On cross-section they were apt to be located next the sarcolemma sheath, and averaged 6.5 nuclei to each fibre, while calculations for normal fibres gave only 1.8 for one specimen and 1.67 for another. At times the nuclei were found in the centre of the fibre, singly or in clumps. They were not so regular or sharply outlined as normal, and seemed larger and plumper. In longitudinal sections they presented long rows of six to twelve to twenty in number, and were generally arranged along the border of the fibre, in lines parallel to the fibre's direction. The connective tissue also showed a moderate increase, but was not to be likened to any inflammatory connective-tissue growth.

This picture was subsequently confirmed by various observers, although certain variations and new peculiarities were noted by many of them. The muscle hypertrophy was seen by all, but no measurements of the enlarged fibres have exceeded those given by Erb for the widest he observed. In addition to the hypertrophy, Dejerine and Sotta, Jacoby,<sup>1</sup> Joseph Koch,<sup>2</sup> and others describe pronounced degenerative changes in the muscles. In Dejerine and Sotta's case, two kinds of muscle changes were noticed, the one affecting the specific tissue of the fibre and the other the indifferent protoplasm, causing the fibres to be more homogeneous and to show the Cohnheim fields more distinctly. In later stages there was a splitting of the fibres and a dissociation of the sarcous elements, which lay in a cloudy, amorphous heap in dilated sarcolemma sheaths. Finally, the resulting loss of muscle tissue was evidenced by vacuoles and empty sarcolemma sheaths. A few examples of solitary atrophy of the fibres were found, and some striated muscle masses were detected without their encircling sheaths. The nuclei were frequently very irregularly situated. Their case is also of interest, as the findings varied in the different muscles examined, being most markedly expressed in the diaphragm muscles and least so in the tongue. They claimed they could trace the progress of the disease in the muscles, and described the first stage as mainly affecting the nuclei, which were greatly increased, and were almost as numerous as they were in other implicated muscles of the body, although the fibres here were not markedly hypertrophied. These variations these authors consider not as the cause, but rather as the result of the disease. They also regard the degree of alterations as somewhat proportional to the severity of the symptoms.

In Jacoby's second paper, published nine years after the first, his views are considerably modified. The hypertrophy is there regarded as due to the abnormal contractility of the muscle after its excision. If this is prevented by proper fixing, immediately after excision, the sections, subsequently cut, approximate closely to normal. One of his muscle sections slipped from its fastenings and confirmed his theory by showing pictures similar to those previously noticed. Consequently, he regards

<sup>1</sup> *Jour. Nerv. and Mental Dis.*, 1887, xiv, 129.

<sup>2</sup> *Virchow's Archiv.*, 1901, cxliii, 380.

the alterations he saw as due to "serous infiltration of the connective tissue and to secondary changes in the parenchyma, thereby nutritionally produced."<sup>1</sup> This accords with Böttiger's criticism of Dejerine and Sotta's interpretation of their muscle findings, for Böttiger claimed the œdema might entirely account for the muscle changes. Koch observed degenerative processes, associated with those of a regenerative nature. Into the interfibrillary spaces, where long rows of nuclei were situated, he noted an ingrowth of capillaries from the perimysium internum. Finally, a sheath would be formed to enclose a new fibre thus cut off. In this way two, three, or even four fibres could be formed from a single one. The degenerative changes resulted in the formation of vacuoles and empty sarcolemma sheaths, and, for the production of the latter, nuclear proliferation was an important factor. In the sarcolemma nuclei he found evidences of an active proliferative process by direct division. A marked increase in the number of nuclei has generally been noted, and some have detected hypertrophy of the individual nuclei, the increase in size being in breadth and not in length. A moderate increase in the connective tissue has not been as constantly found.

The work of Schiefferdecker and Schultze<sup>2</sup> is of special interest, for, in addition to the muscle hypertrophy previously mentioned, they found granulations in the sarcoplasm, which they interpreted as an indication of a specific disease there. They also noted an increase in the number of nuclei, which sometimes appeared in very extensive rows, but no degenerative processes were described or any increase in the number of fibres, such as Koch reported. The granulations were only noticed in those sections of muscle which had been hardened in a solution containing magnesium and sodium sulphate, with sodium chloride and 5 per cent. formalin (Jore's solution). They consisted of small granules, generally circular in shape, and dark black, dark brown, or blackish brown in color, with at times a very glistening appearance. The nuclear changes to Schultze seemed to be primary, and thus to confirm Dejerine and Sotta's theory. They also seemed proportional to the muscle hypertrophy, which did not appear to be functional in origin.

Concerning the *pathogenesis* of the disease much has been written, but it is nevertheless still wrapt in obscurity. (1) The psychopathic theory was first advanced by Thomsen, but it has little clinically to support it, and is now generally abandoned. (2) The neuropathic view attributes the disease to some disturbance of the central nervous system, possibly some functional change in the cord. The theories of Sieligmüller, Peters, Rieder, Westphal, Marie and others can be grouped here. Recently Gregor and Schilder<sup>3</sup> have shown that the myogram obtained by the string galvanometer in this disease proves the muscle contraction is not of purely myogenic origin, but arises from central innervation. (3) The myopathic idea proclaims the muscles to be the primary cause of the malady. Dejerine and Sotta incline strongly to this theory.

<sup>1</sup> *Jour. Nerv. and Mental Dis.*, 1898, xxv, 508.

<sup>2</sup> *Deut. Zeitschr. f. Nervenhe.*, 1903, xxxv, 1.

<sup>3</sup> *Ztschr. f. d. ges. Neurol. u. Psychiat.*, 1913, xvii, 206.

**Symptoms.**—The characteristic symptom is the so-called myotonic disorder, which is revealed when voluntary movements of any sort are attempted. This consists in tonic muscular cramps which forthwith ensue, inhibiting all movements for a short space of time and causing a certain awkwardness and clumsiness of movement, which is very mortifying to the patient. Moreover, increased exertion to conquer this opposition leads only to renewed tonic, muscular contractions. After the same movements have been several times repeated, the muscles, with their antagonists, relax from their state of prolonged contraction, so that the awkwardness disappears, and subsequent similar movements can be executed normally. Occasionally the muscular rigidity is more marked during the second attempted movements. If a few moments of rest intervene, or some sudden obstacle is seen, causing mental disquietude and bringing new muscles into play, the same awkwardness again appears. These tonic spasms or contractions of the muscles are never accompanied by pain, but are sometimes more marked if the patient be fatigued. Cold, heat, depression of mind, and hunger are factors, which may not only bring on the spasms, but make them more severe. At times alcohol and warmth may cause them to lessen. Danillo thought the latter so acted when he gave his patient warm tea, and later proved it by then getting a normal myogram. The spasms may be seen not only in voluntary movements, but also in those of a reflex nature, as in coughing and sneezing, and after mechanical or electrical excitations. Passive movements are unaffected. Erb states the contractions are noteworthy for (1) their slowness, (2) their tonic character, and (3) their continuance after the cessation of the voluntary impulses.

The light forms may cause but slight inconvenience to the patients, who can easily conceal this muscular rigidity by cleverly executed movements. In the severe cases, the patients on attempting to walk appear as if chained to the floor, and the stiffness or rigidity which accompanies each forward movement of the leg must wear off before another step can be taken. With each successive step the stiffness is of shorter duration until it entirely disappears, and the patient may then walk without any further mishap. If, however, he should trip against a stone, turn suddenly around, or bring any muscle, not then in use, into play, rigidity of those muscles will follow, which may result in the patient's falling down. Similarly, in mounting stairs, the first ones seem to offer insurmountable difficulty, but when these are overcome, the rest are easily ascended; in handshaking, the grasp is slow and its relaxation is delayed, while the dynamometer shows the grasp is much below the normal standard; in eating, there may be an inability to close the mouth, when opened, until the spasm of the depressor muscle of the lower jaw relaxes and allows the masseters and internal pterygoids to act; in reading a book, the patient may be unable to raise his eyes for some minutes from the page, although earnestly desiring to do so, and in some cases strabismus is seen, due to spasm of the muscles of the eyeball. Similar difficulties are noted in speaking, dancing, drilling, or any movement requiring voluntary effort.

The muscles of the lower extremity are apt to be most frequently



involved, although those in the upper extremity, as well as the tongue, masseter, eye, and diaphragm muscles, may be affected also. Occasionally the laryngeal muscles are concerned, in which case the speech is altered. Often the same patient shows an implication of the muscles of the extremities and the trunk, but the disease is not equally expressed in all of them, the chief difficulty being in the lower extremities. If the trunk muscles are attacked, all forward movements, such as stooping, throw these muscles into a spasm, but they are unaffected in the movements required in parturition or defecation. Neither are the acts of swallowing, respiration, or micturition interfered with. More rarely, only a few groups of muscles exhibit this malady, as in Oppenheim's patient, in whom the orbicularis palpebrarum alone suffered. The muscles are generally markedly hypertrophied to the naked eye and the patient appears to be of a very athletic build; his strength is below normal and not proportionate to his muscular development. In one of Erb's cases the myotonic disturbance seemed to be more pronounced in the more markedly developed muscles. Later observers have found an atrophy, associated with the hypertrophy, which, in some cases, appeared to be of the progressive muscular type. Bramwell and Addis<sup>1</sup> have found 65 such cases, upon only one of these, however, has an autopsy been performed (Steiner's case)<sup>2</sup>. The muscles of this patient showed a widespread cirrhosis and a tabetic-like degeneration of the posterior columns of the spinal cord. These cases generally present a weakness of the fascial muscles (myopathic face), an atrophy especially of the sterno-cleido-mastoids, the vasti muscles of the thighs and dorsi flexors of the feet, in addition to the myotonic condition. They are of a distinct familial type; Bramwell and Addis, Steinert, Pelz<sup>3</sup> and others believe this is a form of Thomsen's disease, while Batten<sup>4</sup> is the leading exponent of those who would give it a distinct clinical entity. Greenfield,<sup>5</sup> Lange,<sup>6</sup> and Mann<sup>7</sup> have reported instances of myotonia with atrophy associated with early cataract formation in several members of a family.

The presence of von Graefe's sign in exophthalmic goitre was first noted in myotonia congenita by Oppenheim. This association has later been carefully studied by Sedgwick<sup>8</sup> in a family of twenty-nine, in whom it was found in thirteen instances.

The *electrical reactions* have been found as characteristic as the myotonic disorder, and have been collectively designated by Erb "the myotonic reaction." They are as follows: (1) The motor nerves show no increase of irritability to mechanical stimuli; (2) to the faradic current the motor nerves are quantitatively normal, but if the current be strong, the contraction produced on closing the current lasts much longer than it does in health; (3) to the galvanic current, the motor nerves are quantitatively normal; but here, also, if the current be strong, the contraction lasts longer than in health; (4) mechanical stimuli applied to the muscles, as

<sup>1</sup> *Edinburgh Med. Jour.*, 1913, N. S., xi, 21.

<sup>2</sup> *Deutsch. Ztschr. f. Nervenheilk.*, 1909, xxxvii, 58.

<sup>3</sup> *Archiv. f. Psychiat.*, 1907, xlii, 704.

<sup>5</sup> *Rev. Neurol. and Psychiat.*, 1911, ix, 169.

<sup>6</sup> *Deutsch. Zeit. f. Nervenheilk.*, 1910, xl, 65.

<sup>7</sup> *Berl. klin. Wchnschr.*, 1913, l, 846.

<sup>4</sup> *Lancet*, 1909, ii, 1486.

<sup>8</sup> *Am. Jour. Med. Sc.*, 1910, cxi, 80.

by hitting them, induce contractions more early than in health; (5) the faradic current applied directly to the muscles, if strong, sets up a contraction which lasts from five to thirty seconds; (6) when the galvanic current is applied directly to the muscle, K. C. C. and A. C. C. are equally easy to obtain; while in health, as is well known, K. C. C. is more readily elicited than A. C. C. In Thomsen's disease, even with weak currents, the contraction lasts longer than in health; with strong currents it lasts some seconds and relaxes very slowly. With the stable application, well-formed, wave-like contractions are seen to proceed slowly from the cathode to the anode (Hale White). All of the reported cases have not shown an exact resemblance to Erb's cases, which gave the myotonic reaction as Hale White thus described it. They have in almost every instance, though, revealed a normal mechanical, faradic and galvanic excitability of the motor nerves, but an increased mechanical, faradic, and galvanic excitability of the muscles. The peculiar rhythmical, vermicular contractions have only been obtained in occasional cases by the application of a strong and steady galvanic current. Erb, subsequently, did not attach much importance to them, but thought they could be obtained, in every case, by proper manipulations. Babonneix<sup>1</sup> has studied anew the myotonic reaction, and believes it is due to a functional exaltation of the sarcoplasm.

The tendon *reflexes* vary, being increased, normal, or absent. In two of Jacoby's patients the knee-jerks seemed at first exaggerated, but became weaker and weaker, after successive elicitation, until no response was obtainable. The knee-jerks later reappeared after a short interval of rest. There are generally no sensory disturbances, with the exception of rare paresthesia. The quantitative urine examinations have yielded inconstant results.

**Diagnosis.**—The recognition of Thomsen's disease should, as a rule, present no difficulty, as the myotonic reaction and the myotonic disorder are observed in no other malady. Its general hereditary nature will also be of assistance in a diagnosis. Conditions are found, however, which present many forms of resemblance to Thomsen's disease and which may be confused with it. Talma, Furstner, Jolly, and others have described instances showing an abnormal hyper-excitability to mechanical and electrical irritants. Although named myotonia acquisita, they have little in common with the disease we are now discussing, as Jacoby well states, and would be better termed intention spasm. He would apply the term myotonia acquisita to cases closely resembling Thomsen's disease, which come on after injury or overstrain. Another class of cases, presenting temporarily the myotonic disorder and myotonic reaction, he calls myotonia transitoria. Eulenberg's paramyotonia congenita is, also, a closely allied affection, and was noted in one family to the sixth generation. But here the myotonic reaction is absent and the muscular rigidity occurs only after the influence of cold. Delprat found two typical cases of Thomsen's disease, with temporary rigidity of the muscles, in consequence of the effect of cold, in a family with

<sup>1</sup> *Gaz. d. hôp. Par.*, 1913, lxxvi, 2205.

paramyotonia congenita, and Hlawaczek found the two diseases associated in one individual. Myotonia combined with progressive muscular atrophy or hypertrophy has been described, and Schultze, Bettmann, Mill and Kaiser, and others have noted its association with tetany and athetosis. Hoffmann, in one case, and Nalbandoff, in two cases (brothers), have found tabes as an accompanying affection. The former also observed a case with multiple neuritis as an additional complication.<sup>1</sup> In 1907 Pelz considered the atypical cases of Thomsen's disease, and placed paramyotonia congenita in this category. He thinks, in the atypical forms, there are no absolutely pathognomonic symptoms, but considers the disturbances of the mechanical and electrical excitability as the most constant. If the disease develops late in life, he does not regard it as acquired, but as due to an exacerbation or a long latency of a disease which, although present, was slow in revealing itself.

**Prognosis.**—The intensity of the disease varies greatly; in the lightest cases the patients may hide the rigidity of their muscles by cleverly executed movements and experience no great inconvenience; the more severe forms unfit those afflicted for many occupations and cause them to undergo many hardships. It is an essentially chronic disease, which appears to progressively increase up to puberty or even later. In some cases relapses and remissions mark the progress. It is never fatal by itself, and no recovery from it has ever been reported.

**Treatment.**—Nothing which has been hitherto tried in the way of treatment has been of any avail. Hydrotherapy, gymnastic exercises, and massage have given favorable results in the hands of von Bechterew, Rosenbach, and Wersiloff, but nothing permanent has been accomplished. Thomsen thought he, personally, felt better the more active his life was. It appears that something may be done in the way of prophylaxis by cautioning the patient to lead a life as free as possible from mental excitement and injurious influences, such as cold.

### AMYOTONIA CONGENITA: OPPENHEIM'S DISEASE.

**Definition.**—A general or localized flaccidity of the muscles in childhood, which is associated with a weakness or loss of the tendon reflexes. Attention was first called to this condition by Oppenheim in 1900, by whom it was named myatonia congenita. This designation, however, is so similar to myotonia congenita (Thomsen's Disease), that we prefer the above term, to avoid confusion.

**Etiology.**—The disease is observed in children in the first months of life, and is essentially congenital in origin, without any hereditary tendency. Trauma at birth has not been noted in any instance. Seventy cases have now been reported, upon thirteen of which autopsies have been performed. The cause of the disease is unknown.

**Pathology.**—Oppenheim<sup>2</sup> considered the lesions were in the muscles, although he did not entirely exclude an affection of the anterior horn cells

<sup>1</sup> For a full list of the various diseases which have been found associated with Thomsen's Disease, see Ascanzi, *Monatsschr. f. Psychiat. u. Neurol.*, 1912, xxxi, 201.

<sup>2</sup> *Monatsschr. f. Psych. u. Neurol.*, 1900, viii, 232.

as the cause. Since then the same findings have been generally noted, which have been observed by other investigators in the different forms of myopathy. The muscle fibres are mostly reduced in size and atrophied, the muscle nuclei are markedly proliferated and the connective-tissue is much increased. Fat also is observed in the muscles, all of which points either to the defective development of the muscles or to retrogressive changes in previously normal fibres. There are also changes in the nerve tissue, as seen especially in the diminution in number and the vacuolar degeneration of the cells of the anterior horn. Acute swelling has also been described in these cells. The changes in the thymus gland are considered merely coincident. Marburg considered the malady to be due to a fetal poliomyelitis, while Rothmann calls it Werdnig-Hoffman disease, but Oppenheim<sup>1</sup> considers both of these views absolutely untenable. Councilman<sup>2</sup> attributes the disease to a toxin produced in the mother.

**Symptoms.**—The parents of the child so affected are usually the ones to note that the whole body of the child or only the limbs, especially the lower, are flaccid and motionless. This flaccidity may be so marked as to render the joints abnormally movable. Edema is a trophic disorder, which may be present to a very marked degree. There is also a weakness or a loss of the tendon reflexes associated with this condition. Active movements may be impossible or only slightly impaired. The cerebral function is undisturbed. The muscles of the eye are not affected, but the facial and pharyngeal muscles, with those of respiration, may become involved. The implicated muscles are flaccid, soft, and thin, but not atrophic. Collier and Wilson<sup>3</sup> have observed the presence of secondary contractures in some of the affected muscles. The electrical reactions show a quantitative alteration or a complete absence, but in mild cases the reaction is normal. Collier and Wilson described a reduction of the faradic with a retention of the galvanic excitability (amyotonic reaction) but Oppenheim could not corroborate it. No disturbances of sensation are met with and the sensorium is clear. The tendency to relapses should be noted.

**Diagnosis.**—The absence of family tendency as a factor will differentiate this affection from progressive muscular atrophy, while poliomyelitis occurs acutely in a previously healthy child. Amaurotic family idiocy shows a history of a family tendency, different microscopic and ophthalmoscopic findings, and an entirely different clinical history.

**Prognosis.**—The children attacked may show in time some improvement, but the outlook does not appear very encouraging. If the condition is generalized, the respiratory muscles are always attacked. In six instances bronchopneumonia caused the fatal termination.

**Treatment.**—There is no known special treatment. The use of massage, electricity, and nerve tonics is indicated.<sup>4</sup>

<sup>1</sup> *Lehrbuch der Nervenkrankheiten*, Berlin, 1913, 6 ed., 280.

<sup>2</sup> *Am. Jour. Dis. Child.*, 1911, ii, 340.

<sup>3</sup> *Brain*, 1908, xxxii, 1.

<sup>4</sup> For a further consideration one is referred to the following articles: Archangelsky and Abrikossoff, *Arch. f. Kinderh.*, 1911, lvi, 101-129 (Bibliography); and Foot, *Am. Jour. Dis. Child.*, 1913, v, 359.

## CHAPTER XXV.

### ARTHRITIS DEFORMANS.

By THOMAS McCRAE, M.D.

**Introduction.**—It is necessary for anyone writing on the subject of chronic arthritis to state definitely, so far as possible, what conditions are included under various terms and what is meant by the designations employed. It is well to confess that our knowledge of chronic diseases of the joints is in an unsatisfactory condition, and that great difference of opinion exists on many points. The best that one can do at present is to put forward his views in such a form that others can at any rate understand what is meant, and if they disagree, have no difficulty in recognizing where the difference lies. Much of the pathological and clinical material is a matter of record; the conclusions drawn are more or less matters of opinion.

In this article the designation arthritis deformans is used to include the group of cases of arthritis which have as prominent features a tendency to chronicity and to more or less permanent change in the joints or structures about the joints, those forms of arthritis with a definite etiology (*e. g.*, gonorrhœal arthritis) being excluded. That this is a satisfactory classification cannot be held, but it seems the best we can have at present. In gonorrhœal arthritis there may be chronicity with more or less permanent change in the joints, yet we have a definite etiology by which it can be classified. Determine a definite etiology for any case or group of cases and they can be separated from the general class. Taking arthritis generally we can divide the various forms into:

1. Traumatic arthritis.
2. Arthritis due to infection with definite bacteria, such as the gonococcus, streptococcus, tubercle bacillus, pneumococcus, etc., which may be obtained in the joint. It is to this group that the term infectious arthritis properly belongs. It is probable that rheumatic fever belongs here, as the evidence for it being due to infection with a specific organism is very strong.
3. Arthritis occurring in certain diseases, such as scarlet fever and syphilis, and apparently dependent on the cause of the disease, although in some cases it may be due to a secondary infection.
4. Gout.
5. Arthritis due to the injection of various sera.
6. Arthritis secondary to disease of the nervous system, as locomotor ataxia and syringomyelia. In this trauma may play a part.
7. Arthritis in hemophilia, purpura, etc.
8. A large group remains, the cases of which for the present are put together. In this group there is almost an endless variety, many of

them showing acute features at times, others being chronic throughout; the degree of involvement varies greatly, but in all there is the predominant feature of a tendency toward some permanent change in the joint or structures about the joint. In this article these cases are discussed under the heading of arthritis deformans. The designation chronic polyarthritis might be used almost as well as the one chosen.

That there can be any confusion as to the essential nature of the disease as compared with, for example, gonorrhœal or tuberculous arthritis does not seem likely. Suppose we have regarded a patient as having arthritis deformans and subsequently find gonococci in a joint. That case is removed at once to a definite class, just as if we found streptococci or tubercle bacilli. One may say that this group simply consists of those cases with certain forms of chronic arthritic change in which no definite etiological factor can be found. This objection, if it be such, is perfectly correct. When we have a proved etiological factor for all forms of arthritis one kind of classification will be perfectly easy. For the present we are doing what we can to make the best of a bad job. It is important to remember that arthritis is always *secondary* to a process elsewhere; the joint condition is not primary.

Have we in this group one disease or two or more? Many take the latter view. Thus, A. E. Garrod<sup>1</sup> considers that we are dealing with several maladies (*e. g.*, rheumatoid arthritis and osteo-arthritis) and not with a single disease. The question is a difficult one to settle. There is no doubt that certain cases of arthritis show features which seem to suggest distinct entities, but others seem to belong to both groups or to lie between them, showing evidences of both. In all chronic joint changes it is well to keep in mind that the same cause may produce varied results, and the same result may come from different causes. The great variation in the picture presented by arthritis due to a particular organism (*e. g.*, the gonococcus) might easily lead to the severe and mild cases being regarded as different, did not the common cause group them together etiologically. So in arthritis deformans the writer feels that there are so many different grades of change, which merge the one into another, that a classification into distinct diseases is not justified.

*"Infectious Arthritis."*—This term has been used in different ways. Some apply it to certain forms of arthritis which come under the designation of arthritis deformans as given here. These are the cases of arthritis, which occur secondary to some focus of infection, such as tonsillitis, but in which no organisms are found in the joint. Is this designation a good one? The writer thinks not, for various reasons. The term is with propriety applied to such forms as gonococcus or tuberculous arthritis, in which the etiological factor is definitely proved. To use it for cases in which the cause is not established must lead to confusion. Then by some an "infectious" group is separated, that is, on supposed etiological grounds, while other groups, such as atrophic and hypertrophic, are separated on anatomical grounds. This is not meant to imply that those forms occurring secondary to tonsillitis, for example, are not

<sup>1</sup> *A System of Medicine*, Allbutt and Rolleston, revised edition, vol. iii.

due essentially to an infection in some way or other, but the evidence seems about as strong for the atrophic and hypertrophic groups. There is no uniformity in the use of the term, for some use it only for arthritis which is due to a definite infecting organism, while others use it both for this and for the group of cases here described under the heading of the peri-articular form and by some writers termed rheumatoid arthritis. For the cases secondary to a focus of infection, without the presence of organisms in the joints, the terms toxic or toxemic might be used.

By no means all the cases of arthritis occurring with various local infections (*e. g.*, tonsillitis) are to be regarded as belonging to the arthritis deformans group. Many of them are temporary and recovery follows without there being any evident permanent change. It is a question, however, whether repeated attacks of this kind do not always leave some damage. To decide where the line is to be drawn is difficult. The writer feels that if any change is left in a joint the condition probably belongs under the heading of arthritis deformans; the milder cases are better considered as secondary complications of the original condition.

*"Chronic Rheumatism."*—In the view of the writer there is no advantage and every disadvantage in the use of this term. We have suffered and do still suffer from the use of the word "rheumatism." Some employ it as synonymous with arthritis of any form, others for rheumatic fever. If it is used as a synonym for arthritis, then its use for a special form of arthritis should be stopped. It seems much better to use the term for rheumatic fever only. If this be done there is no reason for the term "chronic rheumatism," for there is no evidence known to the writer that rheumatic fever passes into a chronic condition.<sup>1</sup> To apply the term chronic rheumatism to certain ill-defined joint changes is only misleading; call them chronic arthritis and then there is no suggestion that they have anything to do with rheumatic fever. The use of the word "rheumatism" to describe any form of ill-understood pain—muscular, arthritic, synovial, or neural—is a diagnostic sin for which no good word can be said.

**Terminology.**—The designations of a whole or part of this group which have been employed are various, and sometimes it is difficult to know exactly how much is included by any particular one. Among them are: Rheumatic gout (Fuller); chronic rheumatism of the joints (Todd); rheumatic arthritis (Adams); nodosity of the joints (Haygarth); rheumatism chronique primitive (Charcot); goute asthénique primitive (Beauvais); rheumatoid arthritis (Garrod); osteo-arthritis; arthritis sèche; arthritis deformans (Virchow). There is much to be said for the use of the designation "chronic arthritis," as it does not commit one to any view as to the nature of the disease. Against this the criticism may be passed that the process is often acute at its onset or at times during the course. The fact remains, however, that essentially the tendency is toward some degree of chronic change.

<sup>1</sup> Certain cases of rheumatic fever in children may drag on for months, showing one manifestation after another, especially cardiac, and to these the term "chronic" might be applied, but this is not its common use.

**Nature of the Disease.**—To recount the various theories is not necessary. A relationship to rheumatism and gout, an inherited arthritic diathesis, a special kind of degeneration, disturbed nutrition, an inflammatory process, of essential nervous origin, due to injury, reflex irritation, or disease of a supposed joint centre and some form of infection, have all been suggested at some time or other. At present, the view which seems most reasonable is that the disease is dependent on an *infectious* process of some kind. This is not to be interpreted as meaning a definite infection of the joint with organisms. This may occur, but in many cases the arthritis seems to be dependent on local infection in some part of the body, with perhaps absorption of toxins and a secondary arthritis. How far possible absorption of toxins from the intestines may be a factor we are not in a position to state. It seems that in many instances the factors of trauma, wear and tear, and the tendency to degenerative changes in the joints of those advanced in life are additional factors. Changes due to an infectious process, subacute or chronic, may be associated with any of these.

The view of the disease suggested here is that the arthritis is due to an infection of some kind—probably this may be with many different organisms—as a result of which very varied changes may result. The changes involve especially now one and again another part of the joint, with a great variety of results, individual reaction and resistance probably play a large part in determining the changes. As to how far inherited tendencies may affect the process it is difficult to say. Some families seem to have joints which are more susceptible to infection and degeneration.

**Classification.**—Some writers emphasize the distinction between the form in which the bony changes predominate and that in which the changes are more in the cartilage and structures about the joint. That in many cases this is true there can be no doubt, but it does not seem to be true so invariably that it can be used to prove the existence of two diseases. Thus, when one finds a patient showing changes of one form in one joint and of another form in a second joint, the inference is that he has the one disease manifesting itself in different ways. It is possible that two diseases may co-exist, but this is too uncommon to explain the frequency of these cases. Thus, the patient may have marked osteo-arthritic changes in the spine, with distinct hypertrophic changes, and in the peripheral joints, well-marked changes affecting the structures about the joint. Again, in a patient whose predominant changes are peri-articular it may be possible to find distinct hypertrophic changes in some situations. The writer feels that there are so many examples of these mixed cases that they offer a great objection to the adoption of the view that two diseases can be distinguished. That there are striking general differences between typical cases in each group is very true, but are there not just as marked differences between cases in the same group?

For clinical study, it is convenient to make a classification without regarding the forms as distinct diseases. The character of involvement varies, and, as a rule, certain features predominate. This is not always true, and it may be difficult to decide in which group to place a given case. In some of this series it was difficult to decide whether bony changes or



those in the peri-articular tissues predominated. In other cases a single joint showed various changes. In this article certain groups are distinguished for convenience of study, it being understood that the lines between these are not definite, but, as a rule, enable one to put a given case in one of them. Some cases (66) seem to belong to two of them and cannot be definitely classified.

1. A form in which the changes predominate in the structures apart from the bony parts of the joint, although the cartilage is frequently involved to some extent. This group is termed "peri-articular," and in this series 319 cases were regarded as belonging to it.

2. A form in which the chief change consists in marked atrophy both of the bones and cartilages. With this there are usually marked trophic changes in the muscles. This is termed the "atrophic" form, and comprised 9 cases.

3. A form in which hypertrophic changes are the most prominent. There may be marked bony overgrowth, which may occur at the edges of articulating surfaces especially, or in the spine, involving the cartilages and ligaments. This is termed the "hypertrophic" form, and 106 cases belonged to it.

The question arises as to whether one of these forms may pass into another. As a rule, this does not seem to occur, although in some cases the course suggests that cases showing the changes of the peri-articular form may later have those of the atrophic type. Many factors have a possible bearing: atrophy from disuse (which, however, gives a different picture), secondary trophic disturbances, marked general toxemia, etc.

**Etiology.**—The various general factors are as follows:

1. **Sex.**—The figures given by various writers vary greatly, but in the majority of statistics the larger number is said to belong to the female sex. One factor which makes a material difference is whether the cases of spondylitis are included or not. They are usually in males, and if included make the numbers in the two sexes more nearly equal. There is some difference in the relative occurrence of the various forms in the sexes. In the present series of 500 cases,<sup>1</sup> 244 were males and 256 females, cases of spondylitis being included. In a series of 1228 reported cases collected by the Cambridge Committee,<sup>2</sup> it was found that 76.5 per cent. were females and 23.5 per cent. males.

2. **Age.**—It is important to remember that the age of patients when seen may give a false idea of the time of greatest prevalence. The disease is so chronic that many patients come with advanced changes years after the onset. It may be said that the disease can occur at any age except in early infancy. The Cambridge report mentions one case beginning at eighteen months. The more carefully the histories of those in whom apparently the disease begins at an advanced age are studied,

<sup>1</sup> The series used as the basis of this article is composed of the records of patients in the Johns Hopkins Hospital, the Jefferson Hospital, and a number personally observed elsewhere. The report of 110 of them has been published (*Jour. Amer. Med. Assoc.*, 1904, xlii, 1, 94, 161).

<sup>2</sup> *Bulletins of the Committee for the Study of Special Diseases*, Cambridge. This is the report of a special investigation of this disease, and should be studied by all those interested in the subject. It will be referred to as the Cambridge report.

the more often it will be found that there has been arthritis before. Still, the first manifestations may be after the age of eighty years. The records in this series as to the age when the patient came under observation are as follows:

	Number.	Percentage.
1 to 10 years . . . . .	4	0.8
11 " 20 " . . . . .	24	4.8
21 " 30 " . . . . .	84	16.8
31 " 40 " . . . . .	114	22.8
41 " 50 " . . . . .	101	20.2
51 " 60 " . . . . .	98	19.6
61 " 70 " . . . . .	60	12.0
71 " 80 " . . . . .	13	2.6
81 " 90 " . . . . .	2	0.4

The age of *onset* shows that in 40 per cent. this was below the age of thirty years, while only 22 per cent. were below the age of thirty on admission. The figures as to the time of onset are:

	Number.	Percentage.
1 to 10 years . . . . .	21	4.2
11 " 20 " . . . . .	72	14.4
21 " 30 " . . . . .	108	21.6
31 " 40 " . . . . .	107	21.4
41 " 50 " . . . . .	83	16.6
51 " 60 " . . . . .	66	13.2
61 " 70 " . . . . .	27	5.4
71 " 80 " . . . . .	4	0.8
Doubtful . . . . .	12	2.4

The type of disease has some influence as the peri-articular and atrophic forms usually come on earlier than the hypertrophic, which more often occurs in older patients. The changes of this type in the larger joints are especially common in elderly people.

3. **Race.**—This is of special interest in regions where the colored race is a factor. In this series there were 474 whites and 26 colored, which shows a relatively low proportion of colored patients. This is of importance in regard to the influence of certain diseases in the etiology. Thus, the great majority of members of the colored race have had a gonorrhœal infection, and tuberculosis is very common among them. It is interesting to note that they show a higher relative incidence of rheumatic fever than the whites and a much lower relative incidence of arthritis deformans.

4. **Station in Life and Occupation.**—There was no evidence in this series to suggest that any station in life showed a special influence. The patients came from the wealthy and the poor classes in about an equal relative proportion. Nor does occupation seem to have any great influence. Certainly unusual exposure to wet or weather was not marked in this series, and the frequent occurrence among washerwomen, so often commented on by English writers, was not evident. The largest number of the patients lived indoor lives, which raises the question as to whether this may not lower their resistance and predispose to infection, or, at any rate, render them less resistant to a chronic infection once acquired.

5. **Infections.**—With the view now generally held that the changes are due to some form of infection somewhere, the possible causal relation-

ship of previous infections has always to be remembered. Infection occurring shortly before or about the time of onset is important. Of these, some are striking in the close relationship. Six cases of infected wounds are especially remarkable; in none of these was the arthritis suppurative. Boils were associated with the onset in 4 cases, cystitis in 6, dysentery in 8, pelvic infection in 18, otitis media in 10, a pyonephrosis in 1, a miscarriage in 4, and bronchitis in 4.

Of particular diseases, influenza and tonsillitis are of special interest. Influenza had occurred in 28 patients and tonsillitis in 91. The figures for influenza (5.6 per cent.) are lower than in the Cambridge report, in which 9 per cent. was found. In 26 cases there had been frequent attacks of severe pharyngitis, and nasal infection was not infrequent. The tonsillar conditions are usually due to infection with some form of streptococcus, and it is well to recognize that tonsils which are not especially enlarged may contain foci of suppuration in their deeper parts, so that infection from this source may be more frequent than these figures indicate. To gonorrhœa some writers have ascribed importance as an etiological factor. In this series a history was given in 74 cases. If it was an important factor one would expect to find more arthritis deformans among the negroes, few of whom escape gonorrhœa. There was a history of syphilis in 29 cases and of chancroid in one. Scarlet fever, erysipelas, typhoid fever, pleurisy, osteomyelitis, all occurred in a few cases shortly before the onset.

6. **Mouth Infection.**—Pyorrhœa alveolaris, carious teeth, infection about the roots of teeth and under capped teeth are all possible sources of infection and must always be considered. The number of cases in which this source of infection has been proved is steadily increasing.

7. **Trauma.**—In some cases this seems to have an influence, but probably more often in determining the joint in which the symptoms are first evident than the onset of the disease. There is no doubt that in some cases the first manifestations of the disease are wrongly diagnosed as due to trauma. It is often easy for a patient to remember some strain or fall about the time of onset. However, in one form, spondylitis, the influence of trauma seems well established. In other cases the effect of an occupation which throws special stress on particular joints seems to determine the onset or greater severity there. Frequently there is some existing arthritis which is aggravated by the traumatism.

8. **Anxiety and Mental Strain.**—The history of prolonged worry, severe strain and anxiety, troubles of various kinds, etc., is frequently given by the patients (26 per cent. in the Cambridge report). It is often evident that these factors have resulted in lowered health, and this may render them more susceptible to infection, especially if associated with increased work and loss of sleep. In patients who have the disease the influence of these factors in determining recurrence is only too evident.

9. **Family History.**—This is a difficult matter to speak of with certainty. Is there such a thing as an arthritic diathesis? There are many authorities who firmly believe that there is, and yet the matter is difficult of proof. In this series there was a definite family history of arthritis in 114 cases; of these, 30 were apparently arthritis deformans; 30 were

termed "rheumatism," and of these a number were arthritis deformans; 3 were rheumatic fever, and 6 gout, the others being doubtful. Some patients gave a remarkable family history of arthritis deformans. Thus, in one, the father, three brothers, and three sisters all had the disease in a chronic form; one sister escaped. The patient's mother did not have the disease, but her mother and all her brothers and sisters had. The writer knows of one family of six in which every individual had a deforming arthritis of the slow progressive type, some being almost entirely crippled, others having only inconvenience. One patient gave a history of the disease in four generations.

10. **Predisposing Causes.**—While it is often difficult to decide with certainty how much influence these may have, yet in some instances the association is so close that they must be given a place. This is seen not only in regard to the original attack, but also in recurrences. With a patient who has had attacks of the disease, one may be able to predict that with certain events, such as an intercurrent attack of illness, the arthritis is likely to light up. While this is perhaps most marked in the group with peri-articular changes predominating, yet it is also true of those with more marked bony changes. Among these predisposing causes come first those which affect the general health, such as poor hygiene, unsanitary surroundings, hardships, etc. All appear to have an effect. In women the menopause is often of importance, and in 27 of the female patients the disease began about the menopause. This association is seen both in attacks beginning at the menopause and also in women who have had the disease in earlier life, and perhaps almost entirely recovered, in whom the symptoms again become active at this time. Frequent pregnancies seem to have an influence in some patients. The onset may come after frequent childbearing, or, more commonly, pregnancy after the disease is once established may apparently cause a lighting up of the process. The infections also are important. These may be of any kind. Influenza and acute bronchitis, an acute dysentery, or pelvic disease may be quoted as examples. How often exposure, wetting, etc., are factors it is hard to say, but the impression gained from the study of this series is that these are not important. How much importance gastro-intestinal disease may have is difficult to say. It may influence the general health, and so predispose to infection. It must not be thought that predisposing causes are always present. The disease may begin in those who are apparently in the best of health.

11. **Specific Cause.**—The points which support the view that the disease is due to infection are as follows:

(a) *The Character of the Attacks.*—These often suggest an acute infection. The sudden onset, the inflammatory local changes, the fever, the presence of complications, such as endocarditis, pericarditis, and pleurisy, and the tendency to run a certain course are all suggestive. These features are not constant or invariable, neither is the usual picture of any infectious disease.

(b) *The Similarity of the Lesions to those Proved to be Due to Infectious Processes.*—This is seen in the likeness of the lesions in certain joints to those caused by the gonococcus, for example. This is also true of the

bony changes; thus, the picture presented by spondylitis is exactly like that of the spondylitis in typhoid fever.

(c) *The Definite Association between a Focus of Infection and Arthritis Deformans.*—The examples of this are sometimes very striking. Thus, in one patient seen by the writer the deforming arthritis followed a suppurating joint. When this was properly treated the general arthritis improved rapidly, but some deformity was left. Some would say that this was more properly termed "infectious arthritis," but the writer considers all of this group as due to some infection, and this arthritis certainly was deforming. The same association is seen in cases with marked bony changes. In some cases of chronic arthritis associated with bronchiectasis, the joint condition varied with that of the bronchiectasis. The rapid improvement in the arthritis following the removal of a source of infection is often remarkable.

(d) *The Definite Cultural Results.*—In an increasing number of cases organisms are obtained in cultures from various parts, blood, joints, glands, etc. The work of Rosenow is particularly suggestive in this connection.

12. **Nervous System.**—The view was, and is still held, that the cause must be in some change in the nervous system. There is much against this other than the evidence which speaks for an infection. The similarity to the joint lesions in chronic disease of the nervous system is only true of the later stages of some forms of arthritis deformans. Examination of the central nervous system has not shown any changes to account for the arthritis.

13. **Intestinal Infection.**—It seems possible that in some patients the intestine may be the source of a long-continued infection either by the entrance of organisms or by the absorption of toxins. It is possible that the total of repeated slight changes may be due to this. The effect of the continued swallowing of large numbers of streptococci from an infected mouth seems worthy of consideration. As to the possibility of the absorption of toxins from the intestine, one hesitates to attach much importance. Our knowledge of "auto-intoxication" is not satisfactory. One therapeutic point is of interest in this connection: some patients are markedly improved by very free continued purgation. In others, a sharp attack of diarrhoea may result in an immediate gain in the arthritic condition.

14. **Metabolism.**—In the minds of many of the laity and of the profession the cause is supposed to be in some disturbance of metabolism. This is shown by the frequently given opinion that uric acid is the causal agent and by the common reduction in the nitrogenous diet. There does not seem to be any definite evidence in support of this. Is there any evidence that metabolic disturbances are at all responsible? Such disturbances do occur, but are they a cause or a result? There is no evidence to show that nitrogenous metabolism is at fault. It has been suggested that some derangement in carbohydrate digestion may be a factor in some cases. Some patients are undoubtedly made worse by large amounts of carbohydrates, but this seems due to intestinal disturbances. That this may be associated with the absorption of toxins

is a tempting suggestion, but we have no evidence to support it. The most reasonable view seems to be that any metabolic changes are a result and not a cause.

15. **Experimental Arthritis.**—A remarkable instance of the production of "osteo-arthritis" lesions in a rabbit was reported by Poynton and Paine,<sup>1</sup> who obtained in cultures, from the knee-joint of a man with chronic arthritis who died by accident, a diplococcus which injected into rabbits produced an arthritis characterized by destruction and formation of bone and cartilage. This differed from the arthritis produced by the diplococcus of rheumatic fever.

**Pathology.**—In a disease with so many manifestations it is difficult to give any description which will apply generally, and it is well to recognize that there may be all grades of change, from the slightest alteration to complete disorganization of the joint. Nor are the lesions always progressive; there is evidence that slight damage may be done which does not advance. There is no definite relationship between the etiological factor and the resulting change in arthritis; nor is there any regular association between the clinical picture and the pathological condition. Many causes may produce the same result, and one cause may produce various results. We may group the changes as follows:

1. *Effusion.*—This is not constant and shows no peculiar features. The fluid is generally somewhat turbid and contains cells which have no special significance. Early in the course it may be hemorrhagic.

2. *Changes in the synovial membrane,* which are of an inflammatory nature and often hemorrhagic early in the attack.

3. *Changes in the capsule of the joint and surrounding tissue.*

4. *Cartilage.*—This may show erosion, atrophy, and ulceration. In certain cases there may be marked proliferation.

5. *Bone.*—This may show atrophy of slight grade, or in some cases this may be extreme. In other cases there is marked proliferation, which may lead to extreme deformity. The formation of new bone may sometimes occur in the structures outside the joint, as in the ligaments. Atrophy and hypertrophy of bone may occur in the same joint.

Nichols<sup>2</sup> divides the lesions into (1) serous, (2) ulcerative, (3) ankylosing, (4) formative, and (5) fungus or villous. In the Cambridge report the principal changes as made out in the x-ray plates were: (1) Those limited to the soft tissues, (2) dislocation, (3) atrophy or destruction of cartilage, (4) atrophy and destruction of bone, (5) transparent areas, which look like punched-out holes in the plates and contain a gelatinous material; their significance is doubtful, (6) Bruce's nodes, which are small bony deposits, usually on the sides of the phalanges; they are probably not typical of any one condition, (7) deposits of new bone, (8) fibrous ankylosis, and (9) bony ankylosis.

It is convenient to discuss the alteration in the various forms of the disease, although the association of changes may be very marked. Two great forms of change may be recognized: first, those in the soft parts, and secondly, those in the cartilage and bones. In some cases one or

<sup>1</sup> *Tr. Path. Soc. London*, 1902, liii, 221.

<sup>2</sup> *Keen's System of Surgery*, vol. ii.

other of these may predominate, but in many both will be found. It is frequently only by the x-ray plates that the second group may be recognized without opening the joint. Certain points may be noted. One is that suppuration never occurs. Another is that the descriptions of changes found postmortem, years after the acute features have subsided, do not necessarily give any indication of what occurred early in the course. They are the remains of processes long since over. Again, secondary features are common, especially contractures and atrophy of the muscles. This latter is sometimes due largely to disuse, but again must be due to changes associated with the disease, as it comes on too rapidly to be due to disuse, and suggests some trophic disturbance. Again, there may be mechanical results, such as dislocation, of varying degree, and with or without marked changes in the cartilage or bone.

In the peri-articular form there is often a considerable amount of *effusion*, especially in the more acute attacks, which may persist for sometime. The fluid shows no peculiar features, although early in the course it may be distinctly hemorrhagic. Cultures from the fluid are usually negative. The findings in an instance in which the knee-joint was opened may be quoted as characteristic. There were some slight hemorrhages in the neighborhood of the joint, which on being opened was found to contain 100 cc. of fluid. The synovial membrane was injected and showed papillary outgrowths of somewhat translucent connective tissue. The outer condyle of the femur showed erosion of the cartilage, the articular surface consisting of enamel-like bone; the corresponding surface of the tibia was the same. Certain areas were elevated, apparently due to overgrowth of cartilage. Sections of the synovial membrane showed on the surface an exudate of granular material and leukocytes. Below this were many bloodvessels and many leukocytes. Then came a layer of endothelial cells, which merged into the underlying fibroblasts.

**Synovial Membrane.**—This usually shows some thickening, which is more marked in the peri-articular form. If the joint is opened early in the course, the synovial membrane is usually swollen, injected, and redder than normal, the very vascular appearance giving a striking picture. The surface sometimes shows a velvet-like appearance, or is covered by a material not unlike granulation tissue. It may be covered with small processes of varying size or shape. The likeness of these to tubercles has led to that diagnosis being made. The synovial membrane may be thrown into folds. In some cases it becomes adherent to the adjoining cartilage, and where this occurs the cartilage is affected and usually destroyed. Histologically in the early stages there may be a marked hemorrhagic condition. The capillaries are markedly distended, and there may be considerable extravasation into the tissues. Nicholson in the Cambridge report describes two layers, the outer of which is more fibrous than the inner, and in it many fat cells may be seen. There is no marked collection of leukocytes, as is often seen in the inner layer. This is thinner, composed of delicate arcolar tissue, and in some places resembles myxomatous tissue. Sometimes it contains large numbers of fat cells. One interesting feature is the presence of areas containing

large numbers of mononuclear leukocytes, usually around the blood-vessels which show an obliterating endarteritis. There may be marked proliferation of this layer with the formation of villi. In some cases there may be an entire absence of the endothelial lining.

As the process advances there is usually thickening of the synovial membrane and an increase in the fibrous tissue. In some cases a certain amount of necrosis may occur. With proliferation of the synovial membrane the so-called villous arthritis may result, which in some cases is most marked. The villous processes are deep red in color and of very varying shape and size. They sometimes reach such a size that they distend the joint. As a rule, there is comparatively little effusion in these villous cases. Secondary changes may result; there may be calcareous deposits in the membrane itself or portions of the villi may become detached. These subsequently may undergo calcareous change. In some cases the synovial membrane becomes closely adherent to the surface of the cartilage, the destruction of which is most marked at this point. This is well seen in the case reported by Hale White.<sup>1</sup>

In the peri-articular form there is usually a good deal of thickening in the structures about the joints, and the ligaments may be involved. This is especially well brought out by the *x*-ray plates. These may show the changes to be almost entirely outside the joint proper.

**Cartilage.**—The changes are very varied in extent, and the slighter degrees may only be evident on opening the joint or in the *x*-ray plate. In the early stages the cartilage may be injected. The earliest change appears to be erosion, which in some cases seems to be determined by the proximity of the affected synovial membrane. The extent of this varies greatly; it may be very slight, or go on until the whole cartilage has disappeared and the bone is exposed. Histologically there is fibrillation of the matrix, with disappearance of the cells. This may involve only the superficial layers and the structures below are normal. The process may advance until no normal cartilage remains. Apparently this is not always general, and there may be marked loss of cartilage in local areas; in other cases the whole surface shows thinning. The gross appearance of the cartilage is often striking. The surface is injected in parts, and often irregular, owing to the areas of erosion. In other cases the cartilage becomes liquefied and the corpuscles disappear with the formation of cysts filled with mucoid fluid. Nicholson describes the occurrence of necrosis in the cartilage with the formation of non-staining areas. This, however, he regards as being possibly due to gout.

In some cases there is apparently some formation of new tissue, in others there is considerable replacement of cartilage by fibrous tissue. The replacement of cartilage by bone is especially marked in the spine. In the peripheral joints this rarely occurs. The result in any case is likely to be bony ankylosis.

**Bones.**—Here the changes vary with the form of the disease. In the periarticular type there may be comparatively little change, although in severe cases there is considerable atrophy. This, however, is not

<sup>1</sup> *Guy's Hospital Reports*, 1902, lvii, 25.



PLATE XIII



Skiagram showing some thinning of the bones, marked destruction of the carpus, and alterations in the metacarpophalangeal and finger-joints.



PLATE XIV



Skiagram of Knee-joint, showing atrophy of the cartilage.



distinctive and is similar to that seen sometimes with a fracture. If the changes in the cartilage result in its complete destruction the underlying bone becomes exposed and eburnated. If the process is very acute the bone is more vascular than normal and necrosis may result. In some cases the bone becomes rarefied and less dense. This can be seen in the *x*-ray plates and in pathological specimens.

In the so-called *atrophic* form the changes are of a different character. The most striking thing is the marked rarefaction of the bones throughout. With this usually goes destruction of the cartilage, so that as an end result (*e. g.*, in the wrist) there may be entire loss of the divisions between the carpal bones.

In the *hypertrophic* forms there is new bone formation, which may vary greatly in extent. The proliferation is most common at the edges of the articular cartilages, and the ossification may include parts of the adjoining fibrous or ligamentous tissue. This may result in bony outgrowths of varying size and shape. In some joints these projections may cause locking of the joint. In the knee-joint, bony outgrowths may be seen on the patella or at the edges of the femur or tibia. In the hip-joint these may be marked and surround the edge of the acetabulum with an irregular ring of bone. The so-called Heberden's nodes on the terminal phalangeal joints are formed of bone. The same changes are sometimes marked about the metacarpophalangeal joints. In the spine the new formation of bone may replace the cartilages, but more frequently occurs in the spinal ligaments, and the anterior and lateral ligaments may be entirely replaced. In all of these irregularity of the deposits is the rule, and it is rare to find the deposits the same on both sides of the vertebræ. The deposit of bone has been compared to lava which has flowed down and solidified. Wright has well compared the appearance as sometimes being much like a "guttering" candle. It may be noted that spondylitis with deposit of new bone is common in animals, especially in horses.

**Muscles.**—Atrophy is the striking feature, and this is generally found to be spread throughout the muscle. In some cases degeneration has been found, and there is proliferation of the nuclei with increase of the interstitial tissue. At times the changes are probably due to direct extension from the joint, in other cases it is suggested that they are due to trophic influences.

**Nervous System.**—No changes have been found which can be regarded as at all characteristic. In a few instances changes in the anterior horn of the cord have been noted. Vacuolation of the ganglion cells has been associated with altered staining reactions. In the peripheral nerves, neuritis has been found, but this is probably secondary. It is easy to appreciate that in arthritis of the spinal joints the inflammation may involve the nerve roots, or in the hip the sciatic nerve. These changes are apparently always secondary.

**Metabolism.**—No very striking changes have been found. King,<sup>1</sup> in a study of the metabolism, found a mild acidosis, with an increase in

<sup>1</sup> *Johns Hopkins Hospital Bulletin*, 1907, xviii, 274.

the organic phosphates and a retention of calcium, magnesium, and phosphorus. The study of the urine did not suggest intestinal putrefaction. In some cases of the atrophic form the calcium output is much increased.

**Symptoms.**—While an exact classification is difficult, it seems best to discuss the clinical features under the various forms, always having in mind that there are not invariably clear-cut distinctions between them, and that it may be difficult to say to which class a patient belongs.

1. **Peri-articular Form.**—This is the most frequent form, and comprises the cases described as rheumatoid arthritis. The changes are most marked in the synovial membrane and peri-articular tissues. The cartilage is also affected in many cases, but the damage is rarely extreme; whatever change occurs in the bone or cartilage is secondary. In this group we find every grade of change, from that in the patient who has a light attack, resulting in very little thickening about the joint, to the patient who has changes so marked that he is a miserable cripple, perhaps unable to feed himself. There were 319 cases regarded as this form; there were others which were predominantly of this group, but showed features of the hypertrophic form as well. The females outnumbered the males in about the proportion of 3 to 2.

**Onset.**—This is usually in one of two ways, acute in one or many joints or gradual in one joint, others being slowly involved subsequently. A point deserving of emphasis is the suddenness of onset in some cases. It is not unusual for patients to be able to state the exact date of onset; some can even give almost the hour. Sudden disability is sometimes a feature. Thus, in one patient the onset was with severe pain while he was walking on the street. The pain was so intense that he had to be carried home. The onset was acute in about 40 per cent of the cases in this series. The sudden onset is usually with an acute arthritis of many joints, which are red and swollen. Occasionally one joint is suddenly involved acutely for a day or two and then others follow. There is rarely any marked constitutional disturbance or chill.

The *gradual* onset may be in one or several joints. It is often found that one joint becomes swollen or painful, perhaps never red, and after some weeks, or possibly months, another follows. As a rule, the gradual onset is of more serious omen than the acute one.

**Joint First Involved.**—Leaving out the cases in which the onset is with the simultaneous involvement of many joints, many more have the first symptoms in one of the joints of the leg than in the arm. It has been said that the disease is prone to attack first the joints which are most used. However this may be, the statement so often repeated that the disease usually begins in the joints of the hands is certainly not correct. In this series the figures of the first joint affected, when this was definitely known, are as follows: Many joints, 22 per cent.; neck, 3 per cent.; shoulder, 12 per cent.; wrist, 8 per cent.; hand, including the thumb, 10 per cent.; hip, 7 per cent.; knee, 19 per cent.; ankle, 3 per cent., foot, 13 per cent. Certain combinations comprised the remainder.

**Character of the Attacks.**—As in the mode of onset, so in the course, two principal forms stand out, the one with acute attacks of polyarthritis, the

## EXPLANATION OF PLATE XV<sup>1</sup>.

FIG. 1. Metacarpo-phalangeal joint.

FIG. 1 *a*. Photograph of the same joint after maceration.

FIG. 2. Skiagram of a metacarpo-phalangeal joint, showing atrophy of cartilage and formation of new bone at the base of the first phalanx and head of metacarpal; a bony spur is also seen.

FIG. 2 *a*. Photograph of the same joint after maceration; the atrophy of cartilage, eburnation at head of bone, formation of new bone around joint, and a typical bony spur are well shown.

FIG. 3. Skiagram of first phalangeal joint, showing destruction of cartilage and fibrous ankylosis.

FIG. 3 *a*. Photograph of same joint after maceration.

FIG. 4. Skiagram of the wrist, showing bony ankylosis and considerable atrophy of the bones forming the joint.

FIG. 4 *a*. Photograph of the same specimen after maceration. This specimen on dissection showed complete bony ankylosis of the carpi at their articulation, and fibrous ankylosis of the ulnar, radius, and metacarpi.

FIG. 5. Skiagram from metacarpo-phalangeal joints, showing atrophy of cartilage and bone with fibrous ankylosis.

FIG. 5 *a*. Photograph of macerated specimen of the same joint.

FIG. 6. Skiagram of the thumb, showing atrophy of bone and destruction of the cartilage of the metacarpo-phalangeal and phalangeal joints. Well-marked fibrous ankylosis is present.

FIG. 6 *a*. Photograph of the same specimen after maceration.

## EXPLANATION OF PLATE XVI.

FIG. 1. Skiagram of a terminal phalangeal joint, showing atrophy of cartilage, early deposit of new bone in the ligament of joint, and a light area.

FIG. 1 *a*. Photograph of the same joint after maceration. A cavity containing a deposit of urates is shown.

FIG. 2. Skiagram of the first phalangeal joint, showing atrophy of cartilage, formation of new bone around joint, and a well-marked projection from the shaft of the first phalanx.

FIG. 2 *a*. Photograph of the same joint after maceration, showing a deposit of bone around joint and on the shaft of the first phalanx.

FIG. 3. Skiagram of first phalangeal joint, showing atrophy of cartilage and formation of new bone.

FIG. 3 *a*. Photograph of the same joint after maceration.

FIG. 4. Skiagram of wrist, showing ankylosis.

FIG. 4 *a*. Photograph of the same specimen after maceration. A longitudinal section has been made through the carpi and wrist-joint. The carpi are fused into a mass of light cancellous bone. The articulations of the carpi with the ulnar, radius, and metacarpi show fibrous ankylosis.

FIG. 5. Skiagram of metacarpo-phalangeal joint, showing atrophy of cartilage and the formation of bony spurs at the head of the metacarpal bone.

FIG. 5 *a*. Photograph of the same joint after maceration. The bony spurs are well seen.

FIG. 6. Skiagram of a metacarpo-phalangeal joint from a case of arthritis deformans, showing atrophy of cartilage.

FIG. 6 *a*. Skiagram of a corresponding joint from a case of chronic gout, showing similar changes.

<sup>1</sup>The writer wishes to acknowledge the kindness of the Cambridge Committee, especially T. S. P. Strangeways, and the Cambridge Press, in being given permission to reproduce these plates, which are taken from the *Bulletin of the Committee for the Study of Special Diseases*, 1907, vol. 1, Nos. 3 to 9, Cambridge.

# PLATE XV

FIG. 1

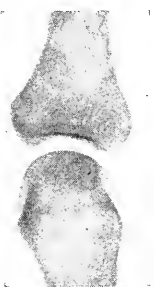


FIG. 1a



FIG. 2



FIG. 2a



FIG. 3



FIG. 4

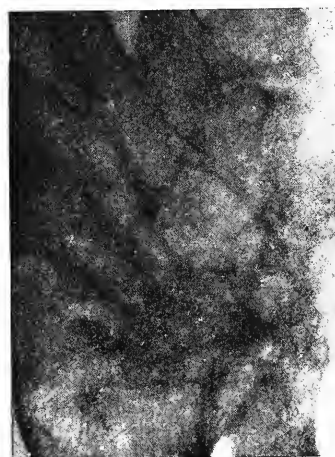


FIG. 4a



FIG. 3a



FIG. 5



FIG. 5a



FIG. 6



FIG. 6a





# PLATE XVI

FIG. 1



FIG. 1a



FIG. 4

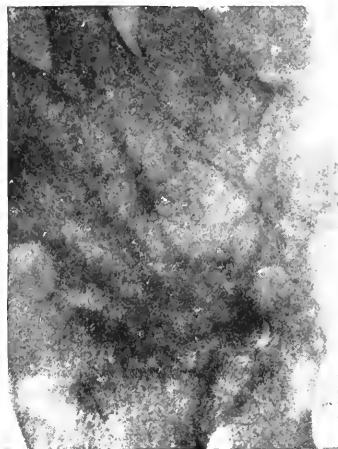


FIG. 2



FIG. 2a



FIG. 4a



FIG. 3



FIG. 3a



FIG. 5



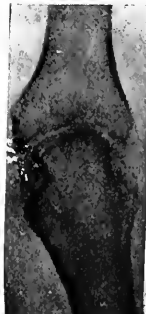
FIG. 5a



FIG. 6



FIG. 6a





other with a slow, chronic, more or less progressive course. There are many combinations and varieties. Thus, there may be repeated acute attacks at intervals, the patient being comparatively well in the meantime, or after the acute attack there may be a slow, gradual progress, or during the course of the chronic form there may be intercurrent acute attacks. However, it aids our clinical knowledge to keep these two classes in mind, recognizing that there are some which cannot be placed in either. In this series about one-third might be classed as acute, one-half as chronic, and the remainder with features of both.

*The Acute Form.*—In many ways this group is much like rheumatic fever, the onset is often sudden, with the involvement of many joints. These are red, swollen, painful, and tender to the touch. There is often effusion in the larger joints. Certain joints not often involved in rheumatic fever may be affected, such as those of the cervical vertebræ, the temporomaxillary and sternoclavicular joints. One point is important—when a joint is once attacked it rarely clears immediately and the involvement is usually for the duration of the attack. Again, as the acute features subside, the joint usually shows some permanent change which may be slight, but it is there. Thus, the finger-joints show slight increase in size or the capsule of the knee-joint is thickened.

Certain general features may be noted. The temperature, as a rule, is not very high, being generally about  $101^{\circ}$  or  $102^{\circ}$  F. Occasionally higher figures are seen, sometimes  $103^{\circ}$  or  $104^{\circ}$ , but this is rarely for longer than a few days. After a time the temperature often drops to  $100^{\circ}$  or  $101^{\circ}$ , and may run along about these figures for some time. Chills and sweating are both uncommon. The pulse generally shows a rate which is comparatively high, and this persists after the temperature has fallen. In a number of cases there is definite *enlargement of the spleen*, which may be made out by palpation or the area of dulness is increased. *Glandular enlargement* is almost always present. This is usually general, with the glands in relation to the affected joints showing the most marked increase in size. As a rule, the administration of the salicylate preparations has little result. The persistence of the arthritis is one of the most striking features.

*The Chronic Form.*—In this the onset is usually gradual, frequently in one joint which swells gradually and is painful. There is rarely any redness, and effusion in the larger joints is not common. In some cases there may be complaint of pain in one joint for weeks before any change can be made out on examination. Other joints are progressively attacked, and it may be a year before there is general arthritis. In other cases one or two joints may be affected for a time, and then with a somewhat acute attack several joints are involved simultaneously. Again, the process may be rather subacute and gradually assume the slow, progressive type. The temperature, as a rule, is not high, often running for weeks from  $99.5^{\circ}$  to  $101^{\circ}$ . It is fairly constant and may never touch normal. The pulse is high, often 110 to 120 or more, and remains persistently so. Enlargement of the *spleen* is not so often found as in the more acute forms. The lymph glands in relation to the affected joints usually show enlargement.

In seeking for an explanation of these various manifestations it is tempting, in the line of a probable infective cause, to consider that the acute type shows a more marked response and more active resistance to the infective agent and its toxins. There is little question but that the outlook, especially for the end, results in the joints, is much better in the acute forms.

*Pain.*—This shows much variation, as in some patients there is comparatively little so long as the joint is at rest; after the subsidence of acute features, pain usually occurs only on movement. In other patients severe pain is constant, and may continue long after all acute features have subsided. The amount of pain and degree of arthritis bear no relationship to each other—in fact, some of the patients who have the most severe pain have very slight joint changes. There are certain situations in which the complaint of persistent pain is common. One of these is about the heel; there may be no special tenderness on pressure, yet every attempt at walking causes so much pain that the patients have to remain at rest. The *x*-ray plates may show no changes to account for it. Pain in the wrist-joint and hand is sometimes very severe and constant. There may be severe pain from an associated neuritis, as occurs especially with involvement of the shoulder- or hip-joint. The pain may follow the distribution of a nerve, and be markedly increased by pressure over it. In the arthritis of the spinal joints the pain may be severe.

*Arthritis.*—In considering this it is convenient to discuss the joints separately, as the features differ considerably. *Vertebræ:* Quite apart from the condition termed spondylitis deformans in this article, we find the joints of the spine not infrequently involved with the peripheral joints. In addition, the manifestations of spondylitis deformans may be found with a general polyarthritis, but in this there are definite changes in the spine. It is important to realize that the joints of the vertebræ may be involved in a general polyarthritis, and apparently no permanent change of any degree be left. There is an acute arthritis which subsides without causing structural change. The most frequent seat of involvement is the joints of the cervical vertebræ. There is complaint of pain, especially on movement; the head may be held more or less fixed, sometimes to one side, as the involvement is not necessarily symmetrical. Pain on movement may be very marked, sometimes more on movement in one direction than in another, and it may be marked on pressure; sometimes distinct grating on movement may be heard by the patient, and it is often audible. There is rarely any permanent fixation, as after a variable period the process subsides and movements are restored. Tenderness on deep pressure may persist for some time.

In addition there may be a similar process elsewhere in the spine, with or without the cervical vertebræ being involved. With an arthritis of the peripheral joints there may be the complaint of severe pain in one part of the spine. This is tender on pressure, the spinal muscles are rigid, and movements cause severe suffering. The duration is often for several weeks. It is rare in these attacks to find signs suggestive of

pressure on the nerve roots or marked muscular atrophy. It is important to realize that this may clear completely without any evidence of permanent change. If the point is looked into it is not rare to get a history of these attacks, and a number of patients with definite bony changes in the spine, give a history of previous attacks of this kind, after which there was apparently complete recovery before the permanent condition appeared.

*Temporomaxillary Joint.*—This is involved much more often in this disease than in any other form of arthritis. The patient complains of pain and difficulty in moving the jaw. The extent of involvement varies greatly; there may be little more than the complaint of pain, or the patient may be unable to open the mouth. Not infrequently the joint on one side only is involved, or one may be much more affected than the other. The process is often temporary. Fortunately the amount of permanent change is usually slight. For a time the patient may be able to open the jaws very slightly, and occasionally teeth have to be removed in order that the patient may be fed.

*Sternoclavicular Joint.*—There is pain, aggravated by any movement of the clavicle, and slight swelling. In rare cases the swelling may be quite marked. As a rule, the arthritis clears completely.

*Shoulder-joint.*—This is frequently involved. In the acute stages there is pain and limitation of motion, especially in elevating the arm above the head and in abduction. There is often tenderness on pressure over the joint, and on anything more than the slightest movement. There is rarely marked effusion; the thickened capsule can often be felt. More or less associated neuritis is not uncommon, the pain being described as running down the arm or up the neck toward the head. As the acute features subside there is often considerable stiffness, crepitus, and thickening of the capsule, with diminished motion. If the process be long continued there is generally marked atrophy of the muscles of the shoulder-girdle. After any attack of moderate severity there is almost sure to be some disability left behind.

*Elbow-joint.*—In the acute attacks the elbow is generally held in a somewhat flexed position, movement is often considerably restricted, and the swelling may extend for some distance above and below the joint. This is often more marked about the elbow than in other joints. Movements other than those of flexion and extension are often diminished. Pain is rarely extreme, but there may be considerable tenderness on pressure. In later stages extension is often limited, but there is rarely a marked degree of thickening left about the joint.

*Wrist-joints.*—The changes here are generally characteristic. As a rule, there is considerable swelling, which may extend above and below the joint, so that the ordinary contour is entirely altered. The swelling frequently has a soft boggy feeling, and there is commonly decidedly less pain than in other joints in proportion to the amount of change. Crepitus is often obtained fairly early. In the subsequent stages the swelling usually subsides, but there is often very definite thickening about the joint, with some restriction of motion. In the early stages, there may be complaint of pain in the wrist-joint, especially on the ulnar

side, sometimes associated with weakness, so that there is a tendency to drop objects, before there are any signs of acute arthritis.

*Hands.*—The changes in the metacarpophalangeal joints are often very striking. There is generally some swelling, redness, and tenderness, especially on motion. This leaves behind some thickening and disability. The degree of involvement in these joints is very variable, and only one knuckle may be involved or all of them on both hands. The condition after the acute features have subsided is generally very characteristic. In some cases there may be subluxation of the joints; one form of change, ulnar deflection, being quite common. This may come on very rapidly in acute cases—thus, in one patient it was marked within one month of an acute onset. A striking feature is atrophy of the interossei muscles. In occasional instances this may be present without any involvement of the joints of the wrist or hand. It is worthy of mention that the hands are not always involved.

*Fingers.*—These are involved in a considerable number of the cases and the changes, as a rule, are very characteristic. In the earlier stages

FIG. 52



Hands showing marked late deformity.

there is usually some swelling, which is often most marked at the first interphalangeal joint. The fingers are often held slightly flexed and full extension may be difficult. With involvement of the fingers and the knuckles it may be impossible for the patient to make a fist. As the acute features subside a very characteristic picture is usually left behind, the thickening about the joint giving the so-called pod-shaped or fusi-form finger. Subsequently there may be quite marked changes in the joints, especially in the way of partial dislocations. A common picture is flexion of the first phalanx, with hyperextension of the terminal ones; but all the joints may be partially flexed, or only the terminal one show hyperextension. Lateral deflection is quite common, especially in the terminal joint. Hyperextension of the terminal phalanges may come on very rapidly, as in one patient in whom this was marked within three weeks of the onset. The thumb joints are often involved.

*Sacro-iliac Joint.*—Apparently this is not often involved in this form of the disease, and it may be difficult to decide whether the arthritis is of this form or hypertrophic.

*Hip-joints.*—As a rule, the changes in this form of the disease are not marked as in the hypertrophic form. During an acute attack of polyarthritis there is complaint of pain in the hip-joint and some pain on movement. On examination little may be made out except slight restriction of movement, especially on abduction and internal rotation. If only one joint be involved the patient may walk so as to favor that side. As a rule, permanent damage of any extent does not result, although at times there may be some fixation with the thigh flexed.

*Knee-joints.*—This is the joint most frequently affected and usually on both sides. In the acute stage the joint is often markedly swollen, red, and tender. Effusion often occurs, and the patella may "float." On palpation there may be considerable tenderness, especially on the inner side, and sometimes over the inner part of the head of the tibia. Tenderness may also be marked in the popliteal space. The joint is commonly held somewhat flexed. As the effusion is absorbed the thickened capsule can usually be felt best on the inner aspect of the joint. If there is much change about the joint, some flexion usually occurs with contraction of the tendons, which later may go on to subluxation. In some very acute cases, and in some chronic ones, in which there is a good deal of muscular atrophy, marked enlargement of the bones may be suggested. This, however, is apparent and not real, being suggested by the atrophy above and below the joint.

*Ankle-joints.*—They often show much the same features as the wrists, considerable puffy swelling extending above and below the joints. There may be considerable stiffness and limitation of motion, which, however, is usually not permanent. The bones of the feet may be involved with the ankle-joint or separately.

*Toes.*—The changes are much like those seen in the fingers, but are usually less marked. In some cases there is the complaint of persistent pain in one or more toe-joints without any evidence of active arthritis. Ankylosis may occur in the great toe-joint.

General features of the arthritis. (a) *Symmetry of the Involvement.*—Much stress was laid on this by the earlier writers, but it is by no means the rule when the figures as to the frequency of the involvement are studied. Thus, in 35 among 126 cases of arthritis of the shoulder the process was unilateral, in 22 of 76 cases of the elbow, and in 24 of 89 cases of the hip. In certain instances the disease may involve one side of the body only, or all the joints of one extremity. It is rare for a single pair of joints, as the knees, to be involved alone.

(b) *Distributions of the Lesions.*—This is very variable, and every combination occurs. In the total number of joints involved the arms exceed the legs, but only by a small amount. If the larger joints are considered, the contrary is the case, and the greater number is in the legs. The frequency with which the hands are involved accounts for the larger total for the upper extremity.

(c) *Variability in the Changes.*—This is seen especially in the acute attacks; which involve many joints. The majority may recover without much change, the brunt of the attack falling on one joint. Both hands may seem to be equally involved at first, and yet one regains almost

complete function, the other showing marked change. Sometimes there seems to be some influence from occupation and special strain on particular joints.

*Muscular Atrophy.*—This is commonly present to some extent, but varies greatly both in its extent and rapidity of development. In some patients, especially in acute attacks, atrophy advances so quickly that some trophic influence seems necessary to account for it. This is usually most marked in the hands, the interossei muscles showing very rapid change. In other cases the progress is slow and disuse may seem to be quite a sufficient explanation. In some patients with repeated acute attacks there may be little change with the first one or two, but each subsequent attack shows more change. About the shoulder-joint the atrophy is sometimes well shown. An interesting point is the occurrence of *muscular twitching*, sometimes seen as a fine fibrillary tremor.

*Temperature.*—Two points stand out in the consideration of this: first, that the fever often shows an elevation much less than might be expected from the degree of arthritis; and second, that it may persist for a long period very slightly above normal. The figures for patients who were under observation long enough to obtain satisfactory records (the patients in whom all acute features had subsided are not considered) showed that in the largest number the temperature averaged from 99° to 100° or 101°. Although at times it was higher, yet this rarely persisted, and many patients go for weeks with a temperature between 99° and 100°. This suggests a chronic low-grade infection. In some cases a rise to higher figures occurs with a complication, such as pleurisy or pericarditis. In a few patients no such explanation could be found. The persistence of slight fever with a relatively high pulse rate is often an important point in diagnosis. It must also be noted that with an acute polyarthritis the temperature sometimes remains practically normal.

*Circulatory System.*—Among 500 cases there was evidence of definite valvular diseases in 40 instances. The lesion was mitral insufficiency in 15 cases, mitral stenosis in 5, combined mitral stenosis and insufficiency in 5, aortic insufficiency in 6, aortic and mitral insufficiency in 6, and aortic insufficiency with mitral stenosis and insufficiency in 3. In about one-quarter of these there was arteriosclerosis, so that the lesions might be regarded as sclerotic; in the remainder they suggested an endocarditis. In 26 cases there was an apical systolic murmur which was not definitely proved to be due to organic valvular disease. In some others there were signs of myocarditis.

Pericarditis occurred in 6 cases under observation and during the course of the acute arthritis. Effusion followed in 3; in one of these tapping was done repeatedly and the fluid injected into guinea-pigs, but with negative results. In 3 cases an acute pleurisy occurred with the pericarditis.

*Pulse.*—This shows an increase in the rate in the majority of cases. In about two-thirds of the cases the rate was over 90. The most frequent range is between 90 and 110, only a few patients having a persistent rate above 120. This increase in rate is true not only of the acute periods, but frequently persists after these have passed, and is also seen in the



slowly advancing forms. When fever is present the pulse rate is usually elevated quite out of proportion to the temperature. In a number of cases there were definite evidences of myocarditis, occasionally with dilatation. The blood-pressure shows no special abnormality.

*Respiratory Tract.*—Bronchitis was present in a few instances, and in some it was suggested that the infection which caused it might stand in some relationship to the arthritis. Lobar pneumonia occurred in several patients during an acute arthritis, in one being associated with a marked exacerbation of the joint condition, which rapidly subsided after the pneumonia was over. The greatest interest, however, is attached to the occurrence of *acute pleurisy* with the arthritis. This occurred in 7 cases, one of which was tuberculous and associated with tuberculous disease of the lung. The others were not tuberculous, as was shown by the negative reaction to tuberculin given after the acute features had subsided. In 3 patients the pleurisy was associated with an acute pericarditis. The importance of this as suggesting some infectious process is evident.

*Glandular Enlargement.*—General enlargement is common and the glands in relation to affected joints are usually enlarged. The degree of enlargement varies greatly, those glands which are in relation to the more acutely affected joints showing the greatest involvement. Thus, the joints of one arm may be involved and those of the other be normal, the glandular enlargement being on one side only, or general glandular enlargement is present, more marked on the affected side. Occasionally the enlarged glands are slightly tender. Histological examination shows only a hyperplasia. In some cases the enlargement is persistent.

*Spleen.*—Among 166 cases with notes as to the size of the spleen, it was found that there was enlargement in 38, in 30 the spleen was palpable, and in 8 the area of dulness was definitely increased. In practically all the cases with splenic enlargement, there was general glandular enlargement. The cases of the so-called Still's disease are included here and will be referred to later. It may be said in a general way that enlargement of the spleen was relatively more common among the younger patients, and in acute than in chronic attacks.

*Subcutaneous Nodules.*<sup>1</sup>—Long regarded as diagnostic of rheumatism, this view can no longer be held, as they may occur without evidence of arthritis of any kind. However, it can be said in the majority of cases they are associated with arthritis, and in the experience of the writer this has been rheumatic fever in children and arthritis deformans in adults. In this series they were found in 17 patients (3.4 per cent.), all being adults. They are generally found about the elbow or wrist, sometimes on a finger, are rather hard, round bodies, between the size of a grain of wheat and a pea, quite frequently movable, and sometimes tender. Their duration is very variable and they may disappear in a few days or persist for weeks. Occasionally patients volunteer the statement that they have been noted to appear, disappear, and reappear. They do not seem to be associated with any severe type of the disease or with any special joint features.

<sup>1</sup> See Fitcher, *Johns Hopkins Hosp. Bull.*, 1895, vi, 133, and Hawthorne, *Rheumatism, Rheumatoid Arthritis, and Subcutaneous Nodules*, London, 1900, J. & A. Churchill.

*Urine.*—This showed no features of moment. In about 20 per cent. of the cases albumin was found, and in rather more than half of these casts were present. Diabetes co-existed in four patients. Estimations of the amount of uric acid were made in a number of cases, but showed no constant departure from normal.

*Blood.*—It may be said that the examination of the blood yields little of importance. The majority of patients have a pale, sallow look, and often suggest a degree of anemia which is not confirmed by the blood count. The average percentage of hemoglobin in 230 cases was about 80; the average red count in 220 cases was 4,800,000, and the average leukocyte count in 220 cases was 8500 per cmm. Leukocyte counts taken during the occurrence of a complication, such as pneumonia, are not included. The leukocyte count in 65 cases was over 10,000 and in 13 cases below 5000 per cmm. It is surprising to find that frequently during an acute polyarthritis the white corpuscles are not increased. The average differential count of the leukocytes in 36 cases showed: Polymorphonuclears 76.3 per cent., small mononuclears 15 per cent., large mononuclears and transitionals 6.3 per cent., eosinophiles 1.6 per cent., and other cells 0.8 per cent. In the case of complications, such as pleurisy, there was a marked polynuclear leukocytosis, which showed no unusual features. It may be said that the blood condition was practically the same in all the forms of the disease. The patient with acute arthritis involving many of the peripheral joints showed practically the same blood condition as the one with the spine alone involved.

*Skin.*—One of the most striking phenomena is the presence of pigmentation, which is especially apt to occur on the face, neck, arms, and hands. It may occur almost at any age, the youngest in this series being nineteen years and the oldest seventy-eight years. There is nothing distinctive about this pigmentation. It may be fairly general over the face and neck or occur in irregular areas; on the hands and arms it more often is seen in smaller, more irregular areas. In some patients there are many small pigmented areas, much like freckles. As a rule, the pigmentation is associated with acute general features, and as these subside the discoloration lessens, sometimes, however, leaving some "staining."

In some cases, especially in those with marked and rapid muscular atrophy, the skin has a curious glossy appearance. This is usually in the hands or feet, but may extend to the arms or legs. In some patients the hands and feet may be very blue. Profuse sweating of the hands and feet is common, and may be a source of great discomfort. This may persist long after acute features have disappeared. In the more chronic forms the skin is often harsh and dry. There may be disturbance of sensation, especially as regards pain, and this may be found to differ on the two sides if the arthritis is asymmetrical. R. L. Jones<sup>1</sup> notes that there may be areas showing sensory changes and sometimes local sweating. Complaint of numbness or tingling, sometimes of severe burning sensation, is not uncommon. Erythema, urticaria or purpura, may occur. Herpes of the lips has been seen occasionally.

<sup>1</sup> *The Lancet*, 1902, ii, 1746.

*Reflexes.*—These very commonly show some alteration, but the findings are by no means constant. The deep reflexes are, as a rule, markedly increased, but may be normal or decreased, although they are rarely absent. In many instances they vary in relation to the joint involvement, and may be different on the two sides of the body or in one extremity. They are usually increased on the diseased side. If there is involvement of the temporomaxillary joint, the jaw-jerk is generally increased. The superficial reflexes are very variable, and may be much increased or greatly diminished. The cremasteric reflex is very variable, sometimes being markedly increased, in others normal, diminished, or absent. Ankle clonus was present in a few cases on both sides and in one case on one side only. Patellar clonus was obtained rarely. In many cases the reflexes were especially exaggerated when there was marked muscular atrophy. R. L. Jones pointed out that there is often a correlation between the affected joints and certain reflexes. Thus if the middle and ring fingers alone are involved, the flexor tendons give a more marked response to tapping than the extensors. The reverse is the case if the thumb and index finger be involved. He points out that there is often marked agreement in the gluteal and plantar reactions. The increase in myotatic variability he regards as of some aid in prognosis as it appears early, and if persistent suggests that the process is still active. Its disappearance is of good omen. Jones lays stress on the association of conditions (*e. g.*, of the plantar and gluteal reflexes), as suggesting some affection of certain spinal segments. In some cases there may be considerable uncertainty of movement, which usually does not amount to tremor.

*Edema* of the feet and legs sometimes occurs, apart from any renal or circulatory lesion. It may be seen in acute attacks or more often in patients with marked lesions in the joints of the legs, especially if there is flexion of the knees with immobility. It is also seen in the patients who are unable to get about and sit all day in a chair.

*Features of the Disease in Children.*—There is much uncertainty as to the nature of chronic arthritis in early life. As Garrod points out, chronic arthritis in childhood is of various kinds. Some rare cases suggest the hypertrophic or osteo-arthritic type which may be the same as that seen in adults. Again, bony changes may occur after hemophilic arthritis, as shown by Bowlby. Garrod refers to rare cases in which chronic arthritis follows acute rheumatic fever, a condition which the writer must confess he has never been able to recognize. Then certain cases of tuberculous polyarthritis in children present curious features, and syphilitic arthritis in childhood may be chronic. However, all these are rare conditions and not so common as the form of polyarthritis in childhood, which has been termed "Still's disease." Special attention was drawn to this by G. F. Still,<sup>1</sup> of London. There are three prominent features: a chronic arthritis characterized by enlargement of the joints, marked general glandular enlargement, and enlargement of the spleen. While different diseases may be included under the term, as has been

<sup>1</sup> *Medico-Chirurgical Transactions*, London, 1897, lxxx, 47. *A System of Medicine* (Allbutt), London, 1897, iii, 102.

suggested, yet the writer is of the opinion, that it is not a distinct entity, but arthritis deformans occurring in childhood. All the cases in this series have been of the peri-articular type; none have been recognized as of the hypertrophic or osteo-arthritic type, although one of the hypertrophic form in this series began at the age of fourteen years. There seems no reason why the hypertrophic form might not occur in earlier years. On the view that these cases are manifestations of arthritis deformans in childhood, they are included in the general statistics given before, but their special features will be considered here.

*Age.*—At what age should we draw the line? From the experience of this series it is impossible to say arbitrarily what patients should be put in the class of "Still's disease." There is a series from four years of age up to eighteen with a gradual lessening of the typical picture in the older patients. In this series one boy, aged eleven years, showed a perfectly typical picture of it. Practically the same condition may be seen in still older children, and enlargement of the spleen and lymph glands is by no means uncommon in later years. In childhood these are undoubtedly likely to be more marked and persistent.

In these cases in childhood the arthritis comes on insidiously and advances slowly. It is of the peri-articular type, the thickening is usually marked, and there may be some limitation of motion. There may be marked muscular atrophy and wasting. Enlargement of the glands is marked; they are discrete and rather hard. Their size often stands in direct relation to the degree of arthritis in the neighboring joints. The spleen is usually felt below the costal margin and is hard, its size varying with the arthritis.

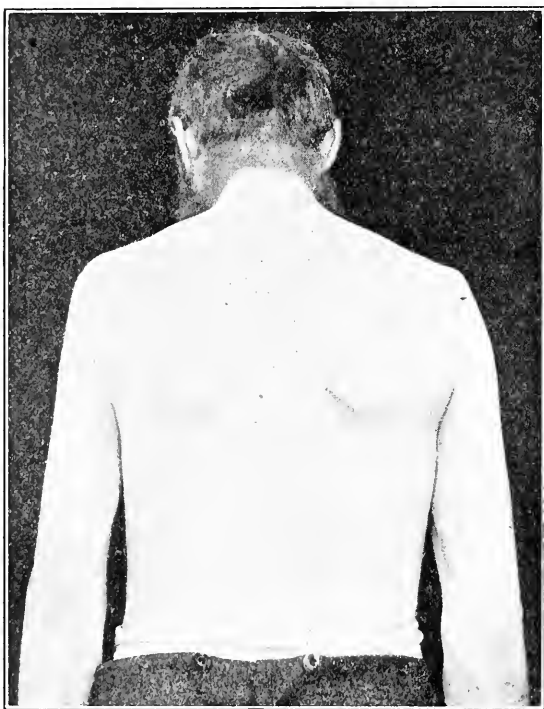
The condition is chronic and in the majority of reported cases has advanced until the patient is completely crippled. This has not been the experience of this series. One patient left somewhat improved, a second left the hospital with the disease apparently arrested and the condition of the joints improving. When seen a year later there had been no return of acute symptoms, the child had grown, was able to get about well, and showed only some slight thickening about the joints. A third patient showed considerable improvement and was able to get about. A fourth, aged eleven years, remained in much the same state.

*The Peri-articular Form in Advanced Life.*—While the view is sometimes expressed that in elderly patients the disease is likely to be more chronic, yet this is not by any means always the case. The first attack may be a very acute one, as in a patient, aged seventy years, who had never suffered from any arthritis previously. Her attack began suddenly, involved many joints, and resulted in considerable permanent change within three months, after which the process subsided. In other instances there may have been a rather chronic progress, or perhaps slight attacks years before, and later in life the first acute attack appears. There is one form occurring usually in stout elderly women which is worthy of note. In such patients the process may be rather chronic, and perhaps has caused slight changes in the small joints, but the brunt of the attack falls on the knee-jerks. These are painful, rather stiff, and apt to "give way" without warning, so that the patients are afraid to walk without

some support. On palpation there may be only slight tenderness and the thickened capsule may be felt. Painter aptly applies the term "knee-sprung" to them. He regards them as being of the hypertrophic type, but some are undoubtedly peri-articular.

*The So-called "Monarticular" Forms.*—This is not a good term, but has been used to describe a form of arthritis usually seen in advanced life, in which one large joint is especially involved. It is a misnomer in the majority of cases, for, if careful examination be made, it is generally found that there is slight involvement elsewhere. The joints most frequently involved are the shoulder and hip. The changes in the

FIG. 53



An example of the so-called monarticular form involving the right shoulder-joint. This patient had slight changes in two fingers, the result of an attack some years before.

shoulder-joint are usually of the peri-articular form, while those in the hip-joint are of the hypertrophic form. The process sometimes attacks the arm which is especially used (*e. g.*, the right arm in a carpenter). The principal symptoms are severe pain, marked disability, especially for certain movements, such as raising the arm and putting it behind the back, and muscular atrophy, often involving the whole shoulder girdle (Fig. 53). Two conditions have to be kept in mind, one is associated with it, and the arthritis is often unrecognized, the other it resembles and may not be suspected. The first is neuritis, which true enough is present, but secondary to the arthritis, and the other is subdeltoid bursitis

As will be pointed out later, one large joint (shoulder or hip) may be involved with the whole spine (spondylitis deformans) but in these cases no special peculiarity is manifested in the peripheral joint, except perhaps marked muscular wasting.

**2. Atrophic Form.**—This is much the least frequent in occurrence, which is fortunate, as it is in many ways the most serious as regards outlook. The most marked anatomical change consists in the atrophy of the bone, which may be very advanced. There is marked loss of the calcium salts. With this there may be extreme disorganization of the joint, which, as regards function, is practically destroyed. The progress in some cases is extremely rapid, and they might be described as fulminating.

This form usually occurs in young adults, and more often in females than in males. It usually begins about the wrist and hand. The symptoms do not differ very greatly from those in the peri-articular form, except that muscular atrophy and disorganization of the joints are more marked. The deformity is often greatly due to dislocation, which is favored by the marked loss of cartilage. Ankylosis, which is usually fibrous, may follow. The hand and wrist have a characteristic appearance. The patient may have difficulty in even lifting the hand or in performing any of the ordinary movements. There may be subluxation of the wrist-joint and marked atrophy of the interossei muscles. The metacarpophalangeal joints may be much relaxed, and there may be great relaxation of the joints of the fingers. The general condition is greatly altered. The patients are emaciated, sallow and anemic, and are very weak. With the progress of the disease, deformities are common. The x-ray plates show atrophy of the bones, quite different from that seen from disuse. With this the cartilage disappears, and the wrist-joint may be represented by little more than a structureless mass. The progress is usually downward, and the outlook in this form is grave.

**3. Hypertrophic Form (Osteo-arthritis).**—In this the changes are more especially in the cartilage and bone, with varying degrees of involvement, but the other structures do not escape and the synovial membrane may be involved. The form is usually polyarticular and apparently may come on at any age, but the majority of patients are older at the time of onset than those belonging to the peri-articular form. In advanced life the lesions are often more suggestive of degenerative than of inflammatory changes, and yet it has to be remembered that the joints are often seen long after the acute changes are over. There were 106 cases belonging to this form in the series. There are two varieties which require special mention, the so-called Heberden's nodes and the spinal form, usually termed spondylitis deformans. The term spondylitis includes many varying conditions. As used here the arthritis of the joints of the spine which occurs without any evidence of permanent bony change—already referred to under the discussion of the peri-articular form—has not been termed spondylitis, but arthritis of the spine. In this section the word *spondylitis* will be used for the cases with definite changes, although really the term includes any inflammatory condition.

Some would doubt the propriety, certainly in many cases, of including spondylitis deformans under arthritis deformans. It is one of the dis-

puted points. The character of the lesions and the frequent association with the undoubted lesions of arthritis deformans in the peripheral joints seem important evidence to the writer. Changes in the vertebræ frequently occur with arthritis elsewhere, and this may be either of the peri-articular or hypertrophic form.

*General Features*—As a rule, the arthritis is polyarticular, the most common exception being the involvement of one hip-joint, the usually termed *morbus coxæ senilis*. As a rule, the age of the patients is greater than in the peri-articular form. Thus, A. G. Garrod found that among 100 cases, 5 were from thirty to forty years of age, 26 from forty to fifty, 42 from fifty to sixty, 21 from sixty to seventy, and 6 over seventy years. Had the cases of *malum coxæ senile* been included the average would have been higher. Yet the disease may come on earlier than shown by these figures. Thus, in one patient of this series, with the onset at the age of fourteen years, the progress was rapid, and in two years he showed well-marked hypertrophic changes in many joints.

As a rule, the general features are not so severe as in the other form. The arthritis is not so acute, and the temperature and pulse are lower. The patient's general condition is much less apt to suffer, and the changes in the joints are much less serious on the whole. The principal effects are more or less mechanical, and muscular atrophy and contractures occur to a much less degree.

*Heberden's Nodes*.—"What are those little hard knobs, about the size of a small pea, which are frequently seen upon the fingers, particularly a little below the top near the joint? They have no connection with the gout, being found in persons who never had it; they continue for life; and being hardly ever attended with pain, or disposed to become sores, are rather unsightly than inconvenient, although they must be some little hindrance to the free use of the fingers." So wrote Heberden under the heading "*Digitorum Nodi*."

These nodes are small bony outgrowths at the terminal phalangeal joints, sometimes covered by a projection of the synovial membrane. They are more common in advanced years, and are found in many people who have not had any evidence of arthritis elsewhere. They may appear in early life, as in one patient, aged sixteen years, in whom the arthritis began two years before, it was of the peri-articular form in the other joints, but he had definite Heberden's nodes of quite large size. They may be the only manifestation of arthritis or occur with a polyarthritis of the peri-articular form or with a spondylitis deformans as the only other manifestation. In some cases they are the first manifestation of a general arthritis. In one patient, a woman, aged thirty-two years, they appeared very rapidly on all the fingers.

They only rarely give rise to symptoms, but some patients complain greatly of the pain, which is usually aggravated by much use or by any injury such as caused by knocking the fingers against some hard object. Numbness, a "dead" feeling or tingling, is sometimes complained of, especially by elderly patients. They are unsightly, and frequently cause disturbance on this account. Their distribution is not necessarily regular on the two hands, from one to eight fingers may be involved, and

in practically any combination. If one finger is especially used in any occupation, as sewing, the nodes on it are likely to be larger than on the other fingers. Occasionally there may be an active arthritis, and the joints are red, swollen, and painful. The onset is sometimes quite acute, and the bony outgrowths appear rapidly. The nodes can be readily felt, and stand out markedly in the x-ray pictures. Sometimes deflection of the end joint of the finger may be caused by them, although this is not common. They often cause some flexion of the end phalanx, and may interfere considerably with the finer movements of the fingers. Garrod points out that small transparent cystic swellings, which he regards as probably being herniæ of the synovial membranes, are sometimes seen on the posterolateral aspects of the joints.

The question often arises as to the significance of these nodes. Many patients who have them carry the opinion that they are a manifestation of the effects of "uric acid." In rare cases they occur in patients with gout, but as a rule, they may be regarded as almost invariably a manifestation of arthritis deformans.

*Thumb.*—The carpometacarpal joint is not infrequently attacked. It may be said that involvement of the thumb-joint is more common in arthritis deformans than in any other form of arthritis. There may be a good deal of bony enlargement, with well-marked crepitus, and more complaint of pain in this than in any other joint. The swelling may be quite marked.

*Metacarpophalangeal Joints.*—These may show marked bony outgrowths, which sometimes are very prominent and project. If with this the joints are much relaxed, great deformity may result.

*Hip-joints.*—While the involvement of this joint is especially frequent and may be the only manifestation of the disease, yet it may be part of a general arthritis. The more carefully the histories are studied in the cases regarded as monarticular, the more often it will be found that there were previous attacks of acute polyarthritis, which possibly have left but little damage, and the more thoroughly the patients are examined, the more often will slight changes be found in other joints. This may be in the form of Heberden's nodes, or perhaps something in a shoulder- or finger-joint. Still, in a great number the principal change is in the hip-joint. As a rule, only one hip-joint is involved.

This form occurs usually in advanced life, although examples in early life are seen. It is much more common in males, and a history of trauma is frequently obtained, although there is often the suspicion that this is secondary to the onset and determined the first marked manifestation. The most marked features are *pain* and *disability*. Pain is not constant, but may be very severe. It may be referred to the joint itself or to the groin, or be especially felt in the leg, sometimes along the course of the sciatic nerve and sometimes being referred to the knee or ankle. This sometimes leads to the diagnosis of sciatica, and it has to be remembered that a neuritis may be present secondary to the arthritis, exactly as in the shoulder-joint. Disability may be marked, with definite limitation of motion. The patients often have a characteristic gait, saving the affected leg, perhaps with tilting of the pelvis. Sometimes walking



is almost impossible, and going up steps is especially likely to cause pain. The patients get up from a sitting posture slowly, and may have difficulty in crossing the affected leg over the other, perhaps having to use the hands to accomplish this. On movement of the leg there may be severe pain and marked grating. Limitation of motion is especially marked on abduction and inward rotation. Extreme flexion may be impossible. There may be shortening of the leg in advanced stages, but in the early stages it is usually only apparent.

There is commonly some muscular atrophy, which may be marked, and is found principally in the gluteal region, which may show distinct flattening. The knee-jerk of the affected leg is usually increased.

*Knee-joint.*—This may be involved as part of a more or less general arthritis, but not infrequently this joint suffers more and may be the most severely attacked. There is usually the complaint of pain and stiffness, with difficulty in going up stairs, kneeling or rising from a sitting to a standing position; a feeling of insecurity with a giving way of the knee is not uncommon. The joint rarely shows much swelling, and in the early stages there is not much restriction of motion. On movements of the joint a loud grating noise may be produced, which Garrod well terms a "scrunch." In the early stages, and some patients do not progress beyond this, it may not be possible to determine that hypertrophic changes are present without an *x*-ray examination. Quite often bony outgrowths from the patella are the first to be recognized by palpation. With progress of the malady, the bony outgrowths become more prominent and may be felt about the femur and tibia. With this there is greater disability and restriction of motion.

The results of the *x*-ray examination suggest that a number of instances of moderate involvement of the knee-joints, seen especially in elderly patients, are of the hypertrophic type. There is but slight arthritis elsewhere, and the process may never advance very far.

*Sacro-iliac Joint.*—This may be involved, although it may be difficult to determine exactly how much change there is in the joint. There is the complaint of pain, which may be referred away from the articulation.

**Spondylitis Deformans.**—This is sometimes termed "poker-spine, poker-back, or rigidity of the spine," and deserves special attention on account of its frequency and importance. As already noted under the discussion of the polyarticular form, involvement of the joints of the spine may occur without, so far as can be determined, any permanent change remaining. These may be termed spondylitis, but hardly spondylitis deformans, so that here the designation is applied to those cases in which there is evidence of definite structural change.

This condition in the spine may occur as the only manifestation, or may be associated with arthritis of the peripheral joints. Of the 120 cases of spondylitis deformans in this series, in 60 the spine alone was involved, and in 60 this was associated with arthritis elsewhere. While the view is taken here that these changes in the spine are manifestations of arthritis deformans, yet this is by no means the universal opinion, and there has been much controversy regarding both the place of the condition in a general classification and as to whether there are really

distinct entities included. Thus, Bechterew has described under the name "Steifigkeit der Wirbelsäule" what he regards as a distinct entity. Strümpell, under the name of "Chronisch ankylosierende Entzündung der Wirbelsäule," and Marie, under the term "Spondylose Rhizomélitique," have designated forms with somewhat different manifestations. An article by Rhein<sup>1</sup> gives a summary of much of the literature. The question as to the identity of the various forms is difficult to decide, but when we remember how diverse the manifestations of the disease are in other joints, it seems reasonable to expect that the same should be true of the spine. The extent of spinal involvement is very variable; in the lower spine the process is frequently most marked on one side, in the cervical region it is much more often on both sides. One patient has the spine alone involved, and then the spine and one hip; another the spine and both shoulders and both hips; one patient has marked signs of pressure over the nerve roots, another has practically none; one patient has kyphosis, another has none; all these variations are found in such a varying degree that to distinguish various forms from their presence or absence seems a mistake. The writer feels that these descriptions are of differing pictures of the manifestations of arthritis deformans.

In the study of the symptoms it is well to have in mind the lesions which are present. These involve an arthritis of the spinal joints, changes in them which are frequently associated with proliferation of bone, atrophy of cartilage with its replacement by bone, and osseous changes in the ligaments. These last are involved frequently, perhaps the anterior lateral ligament most often of all. The extent varies greatly, and the process may extend along several vertebræ, making practically a solid column for some distance, or a process of one vertebra may be joined to that of the next one. The transverse processes are not infrequently involved. On account of the anatomical relations, there is frequently some affection of the nerve roots either through extension of the inflammation by contiguity or by pressure. Following this there are symptoms in the distribution of these nerves. Clinically the results of these changes are seen in many ways. Thus, the arthritis causes pain and muscle spasm; if the articulations of the ribs with the vertebræ are involved there is severe pain on breathing, and perhaps almost entire absence of thoracic respiration. The formation of new bone may interfere with the spinal movements, and we have more or less rigidity as a result. If there is rapid destruction of the intervertebral cartilages without their being replaced by bone we have marked bowing of the spine; if there is rapid ankylosis and replacement of the cartilage by bone, the spine is fixed in a straight position, the so-called "poker-spine." The osseous changes in the ligaments give rigidity and limitation of motion. Pressure on or involvement of the nerve roots may give marked sensory disturbances; thus there may be severe abdominal pain or pain in the distribution of the sciatic nerve. When it is also remembered that the process is usually not symmetrical and may involve

<sup>1</sup> *Jour. Am. Med. Assn.*, 1908, li, 462.

all or only a part of the spine, the possibilities for great variation are very evident. This should be kept in mind in the study of the symptoms.

As to the *etiology*, the most reasonable view is that it is due in some way to an infectious process. The changes are exactly like those which have been found in some cases of the so-called typhoid spine. It has long been known that there may be a spondylitis in association with gonorrhœa. Then in some cases the association with some focus of infection may be very definite. Thus, a tonsillitis or an infection of one of the nasal fossæ apparently stands in direct etiological relationship, and with proper treatment of this the spondylitis promptly subsides. The importance of trauma must be noted; sometimes this accentuates the symptoms, but in other cases undoubtedly determines their onset.

*Occurrence.*—This is probably much more frequent than has generally been supposed. In this series there were 120 cases, of which 60 were spondylitis alone and 60 spondylitis with involvement of the peripheral joints. As regards sex, the figures are striking, for it occurs in the great majority of instances in males. As already noted, it makes a great difference in the relative figures for the sexes in arthritis deformans generally as to whether or not these cases of spondylitis are included. Of the 120 cases here, 97 were males and 23 females. Of the cases in females, the great majority were spondylitis with involvement of the peripheral joints. As to *color* 113 were white and 7 colored. This is a very striking difference, and very difficult to explain. The colored race are just as susceptible to infection, perhaps more so, and trauma must be as common in them as in the whites.

*Age.*—There was no special difference in the pure spondylitis cases and in those with other joints involved. The age at onset in the cases in which this could be determined was:

11 to 20 years	9
21 " 30 "	29
31 " 40 "	34
41 " 50 "	17
51 " 60 "	16
61 " 70 "	8

The time of onset does not differ greatly from that seen in arthritis deformans generally, although the average is at a slightly earlier age. It is worthy of note that the patients with the onset after the age of sixty years were all examples of spondylitis associated with changes in the peripheral joints, and in all of these the onset was fairly acute. As a rule, the onset was gradual. Trauma, usually described as "strain" while lifting, was associated with the onset in 14 cases.

The *complaint* made by the patient is of interest. The largest number complained of pain in the back, but in many there was complaint of pain in the legs, abdomen, or thorax. Several made the complaint of "sciatica." It is well to keep this fact in mind, as there may be no mention whatever of symptoms referred to the back, and attention is easily directed elsewhere. The first symptom varies greatly. In the majority it was pain, referred to the back, hips, or legs, while in others

it was referred to the thorax or abdomen. Several complained of stiffness or weakness in the back. Cramps in the legs were the first manifestation in some patients. All of these may be on one or both sides of the body, but in the majority they are more marked on one side than on the other, especially when pain referred to the legs. The variability in the symptoms is understood if one keeps in mind the anatomical condition.

Throughout the course *pain* is the most marked *symptom*. This may practically be situated anywhere in the trunk or extremities, but the much greater frequency with which the lower spine is involved explains the corresponding frequency in the pain being referred to the legs. It may be in the spine, in the course of certain nerves, or in both situations. In some cases the pain may come on in severe attacks, which make the patient cry out. One feature is the onset of pain in the night after the patient has been asleep. This is most frequent when the disease is in the lower spine. It is exceedingly common for the nervous condition of the patient to suffer, for as a result of the wearing pain and disturbed nights the general health is much affected and the patient becomes very neurotic and upset. As a result the nutrition is affected and the patient may lose a great deal of weight. The symptoms may vary a good deal at different times. Some patients are fairly comfortable as long as they are quiet, but any movement stirs up the pain. Others make a special complaint of pain during the night, which may be more or less continuous, or wakens them from sleep. This is probably due to the muscles being relaxed, so that sudden movement causes pain.

*Examination.*—In discussing conditions which are found one must always remember the great variability, as there are all grades between the patient who has rigidity of the whole spine and the one who has involvement of only two or three vertebræ. The general appearance is sometimes striking. The patient often looks worn, haggard, and sometimes emaciated. The attitude either on standing or walking may be characteristic. If there is general involvement with flexion of the spine the upper part of the body is bent forward, the head projecting, and the eyes fixed on the ground so that the patient has difficulty in looking up. In cases in which fixation has occurred with the spine perfectly straight, the so-called poker back, the patient walks unusually erect. In the cases with limited involvement of the lower spine the patient may limp, and when he stands hold the leg of the affected side with the knee bent. With this there may be distinct tilting of the pelvis. The attitude when the patient is sitting down or getting up is often suggestive. They do these actions slowly, perhaps resting the hands on the thighs, making every effort to avoid bending the spine. If they are asked to stoop and pick up an object from the floor the position is generally very characteristic. They often lower the body by bending the legs and reaching down with the hand, every effort being made to avoid bending the spine.

On examination of the back much or little may be found. The cases of general involvement are so characteristic that they require little description. It may be found that the natural curves of the spine have disappeared, and especially that the lumbar curve is obliterated. In acute cases the condition of the muscles on either side of the spine is

sometimes striking. They are markedly contracted and may stand out prominently. This is most striking when the process is especially on one side, the difference in the state of the muscles being generally very pronounced. The important point is the observation of the movements of the spine, for some or all of these movements may cause pain, and limitation of motion may be very evident. In the early stages this is rarely equal in all directions; with general fixation it may be. Some patients on attempting to bend forward move somewhat to one side. In bending to the sides it may be found that the freest motion is in different parts of the spine on going to the right and left. Sometimes contraction of the muscles becomes much more evident with the movements. In some instances there may be prominence of the spines of one or two vertebræ. Occasionally there is distinct tenderness on palpation over particular spines. In cases in which the vertebræ of the neck are involved it may be possible to feel the bony thickening. If this has been especially on one side the head is rigidly fixed in a bent position. There may be involvement of the sacro-iliac joints, and this usually renders the diagnosis difficult from local involvement of the lower spine.

*Associated Features.*—The frequent complaint of pain in parts other than the back has been noted. This is usually referred to the area of distribution of the nerves, although in some cases there is pain referred to the course of a nerve associated with tenderness on pressure over it. This may especially concern the sciatic nerve, and the ordinary features of sciatica are found. Muscular wasting is quite frequent. This sometimes involves the muscles of the back, but more often those of the leg; thus, there may be marked flattening of one gluteal region, and also wasting of the thigh muscles and these of the leg. As a rule, the reflexes, both the knee-jerks and the tendo Achillis reflex, are markedly increased but this is not invariable. Ankle clonus was obtained in a few instances. With the process in the spine largely on one side, muscular wasting and increase in the reflexes may be confined to the leg of the affected side. Disturbances of sensation are common, and there may be paresthesia or anesthesia. The sensory changes may vary from time to time. Irregular jerking movements of the muscles are sometimes seen.

Features in the *thorax* are often present if the dorsal vertebræ are involved. Thus, in acute stages with involvement of the articulations between the ribs and vertebræ, pain on breathing may be so severe that the patient makes every effort to spare the thorax and the respiration may be entirely abdominal. It may be noted that the same picture is presented in the cases of general arthritis, with involvement of the joints of the spine. In all these forms as the arthritis subsides, and if there is no fixation, the thoracic movements may be entirely restored. In other cases, however, there may be fixation, which is permanent. If this be associated with bending of the spine the change in the shape of the thorax may be marked, and the costal margin may be near the iliac crests.

*X-rays.*—The study of the x-ray plates is of great importance; perhaps the most striking point is the detection of deposits of new bone. These in the majority are extremely irregular, and may be largely on

one side, or especially in the anterior ligaments. This irregularity is really the rule; the symptoms suggest, the x-ray plates confirm it, as does also the study of museum specimens.

*Diagnosis.*—The cases of general involvement are so characteristic that a glance is usually enough to give the diagnosis, but with the local forms the story is different. To J. E. Goldthwait,<sup>1</sup> of Boston, much of the credit of drawing attention to these conditions is due.

The symptoms complained of most commonly are stiffness, interference with motion, pain, and muscular weakness. It may be that many of the indefinite pains complained of in the back, especially in the morning on waking, are due to this condition. "Lumbago" is at times due to this and many of the cases of so-called "sciatica" are caused by arthritis of the spine, and also instances of obscure pains in the legs and about the body. There is no intention of suggesting that every case of pain in the back or of sciatica is due to spondylitis, but in every such case it is most important to examine the back. Some cases otherwise regarded as neurasthenia with complaint of pain in the back will be found to have local arthritis of the spine.

Of the methods of diagnosis first comes the examination of the spine. The patient should be completely stripped, or down to the hips at least. In the majority of cases little is to be made out on inspection. There may be some curvature, or a projecting spine, or more commonly a obliteration of the lumbar curve. Some wasting of the dorsal muscles, or in the gluteal region, or of one or both legs, may be found. In some the attitude is suggestive. They stand with one leg a little flexed and "favor" that side. Next comes the investigation of the mobility of the spine. Any limitation of motion is very readily recognized. This may be equally marked in all directions, or only in one, although usually at least two are combined. He may bend forward but a very short distance, and to one side much less than the other, or lateral movement to one side only may be much restricted. These movements may cause pain, either local or referred. An attempt to bend to the side may cause the same pain in the leg of which the patient complains. The attitude on attempting to pick up an object on the floor is often very characteristic. With this the sensation should be tested; areas of altered sensation are important. The reflexes are usually increased.

The *diagnosis* may be fairly clear from the examination, but there are two important aids—namely, tuberculin and the x-ray picture. The use of tuberculin excludes the most common source of difficulty, although tuberculous disease of the spine rarely gives the same picture as spondylitis. In some instances there is marked involvement of the hip-joints with the spondylitis, and this may have led to a suspicion of tuberculous hip-joint disease. The radiograph, if it shows anything, is usually characteristic. Should the diagnosis not be certain with any of these methods, one help remains—the therapeutic test. Put a light plaster jacket on the patient, which should extend from the axillæ to the level of the trochanters, and if there is spondylitis there should be marked

<sup>1</sup> *Boston Med. and Surg. Jour.*, 1899, vol. cxii, and 1902, vol. cxlvi.

improvement in the symptoms in a few days. This is especially useful in the cases with sciatica. The possibility of sacro-iliac joint disease should be kept in mind in obscure cases.

**Villous Arthritis.**—This is not a distinct entity, but the term is used to describe a condition which may occur from many causes: Trauma, either direct or indirect from flat foot, etc., or from changes in the joint itself, as detachment of a semilunar cartilage; loose bodies in the joint; tuberculosis; syphilis; various forms of infectious arthritis (*e. g.*, gonorrhœal), and not rarely from arthritis deformans. When it occurs as one of the manifestations of arthritis deformans, it has to be regarded as only one part of the general picture.

The degree of proliferation of the fringes varies from a delicate fringe of membrane to large, irregular masses which may occupy the greater part of the joint cavity. They are usually very vascular, and show both fibrous and fatty tissue. The knees, shoulder, ankle, and hip are most often involved, the knee being by far the most frequent. In cases in which the condition followed trauma, it is usually monarticular, but when with general disease it may be polyarticular. The symptoms vary with the degree of change. When the fringes are soft they may cause but few symptoms. In the average case there is discomfort in the joint or pain which is rendered worse by use. There is usually considerable general swelling, which suggests marked effusion, and may cause error unless a careful examination is made. The patella may seem to float and fluctuation may be simulated. Tenderness on pressure may be present. Until one is familiar with the condition, the diagnosis of effusion seems the most likely one; but there is a curious feeling on palpation, very suggestive of a villous arthritis.

**Association of Arthritis Deformans with Other Diseases.**—The relationship between infective processes of various kinds and these joint conditions has been noted. An interesting association is with Raynaud's disease and scleroderma, or both. In this series there were eleven patients in whom scleroderma co-existed with the arthritis, and in five of these there was also Raynaud's disease. It is rather striking that in no instance did Raynaud's disease occur alone, but always with scleroderma. In some instances the scleroderma was present first, but in some instances it and the arthritis appeared about the same time and in others the arthritis antedated the scleroderma. Of the etiology of these diseases we have little accurate information, although it is strongly suggested that they are trophic disorders. It is quite possible that the infection which determined the arthritis may also influence the nervous system. In these cases there is also evidence of trophic disturbances in the glossy condition of the skin, and sometimes in the state of the nails. It was rather striking that the examples of these combinations occurred in women, and nearly all of them were patients of a marked neurotic character, it being necessary to keep in mind, however, that the state of the nervous system is largely secondary to the disturbance of the arthritis. As a rule, the outlook does not seem to be good in this group. The scleroderma seems to have a marked tendency to progress.

In the Cambridge report attention is drawn to a group of cases which

show lesions suggesting both gout and arthritis deformans. These are puzzling and their exact nature is not decided.

**Diagnosis.**—It may be thought that the question of diagnosis would be a difficult one in a disease about which there is so much confusion and which may really include various diseases. Yet actually when the problem is faced the difficulties are not so great. They may be regarded as generally belonging to one of two classes: (*a*) in which the condition is acute and the deforming changes are not greatly in evidence, and (*b*) in which chronic changes have appeared. In the first group it may be difficult to form a definite opinion for some time; in the second we know that the number of diseases causing deformity of the joints is not so large that differentiation is often difficult.

Certain points may be mentioned. That a thorough examination is important seems unnecessary to say, and yet the diagnosis of an arthritis in the larger joints may be cleared up by finding slight changes from a previous attack, perhaps in one finger-joint. An arthritis which recurs and which has left permanent damage in a previous attack is usually arthritis deformans, gout being excluded. Then it is important to learn that this disease may begin suddenly as an acute polyarthritis, which too often is regarded as excluding it. The character of the fever curve, the rapid pulse, the special joint changes, are all aids. In the more chronic forms the diagnosis is rarely in doubt, but unfortunately at this time the benefit of early diagnosis is gone.

Certain characteristics of the arthritis are important. Perhaps the most marked is that when a joint is attacked the condition rarely clears up except very slowly, and there is always a tendency to some change. If it be kept in mind that rheumatic fever does not leave any permanent change in a joint, one frequent source of error will be avoided. Location in certain joints is often suggestive. Thus, to have involvement of the joints of the cervical vertebrae, the temporomaxillary-joint or the thumb-joint, in association with arthritis elsewhere, is often suggestive of arthritis deformans. Involvement of one or two finger-joints is likewise suspicious. Permanent changes in the joints may be found early; for example, the capsule of the knee-joint may show definite thickening, or there may be distinct thickening about a finger-joint soon after the onset. The greatest aid is given by the *x*-ray examination, which should always be made in a doubtful case.

In distinguishing this disease from others, it is convenient to consider, first, the more acute forms, remembering that what is to be a chronic arthritis may have acute features at onset, and, secondly, the more chronic forms. Among the acute diseases are the following:

1. **Rheumatic Fever.**—It may be quite impossible at first to distinguish this. The usual mistake is to regard an acute form of arthritis deformans as rheumatic fever, rather than the reverse. Knowledge of how often arthritis deformans begins suddenly as a polyarthritis with acute features is an aid. There is rarely any shifting from joint to joint in arthritis deformans, and a joint once attacked stays affected. The character of the arthritis is different, as a rule. The joints are usually not so tender to the touch as in rheumatic fever, there is rarely the same degree of



redness, and it can sometimes be made out that the swelling is more in the surrounding tissue than in the joint itself. Certain joints, especially those of the neck, the temporomaxillary joint, and those of the thumb, are rarely attacked in rheumatic fever. The involvement of the finger-joints is often very characteristic. The temperature in arthritis deformans is usually not as high as in rheumatic fever, the degree of arthritis being considered, while the pulse rate may be much higher relatively than the fever. The enlargement of the associated lymph glands is usually greater than in rheumatic fever. Rapid muscular atrophy, especially of the interossei muscles, is suggestive, as it is more likely to occur with arthritis deformans. The lack of response to treatment with the salicylates may excite suspicion. In a doubtful case the first clue is usually given by the persistence of changes in the joints. This should always suggest a reconsideration if the diagnosis of rheumatic fever has been made. In all cases of acute polyarthritis in which there is some question as to the nature, the diagnosis of rheumatic fever should be the last to be made and not the first, as is usually the case.

2. **Gonorrhœal Arthritis.**—This undoubtedly is the most difficult disease to distinguish. Keeping in mind the possibility and a thorough search for gonococci are the great safeguards. The clinical features are much the same as in arthritis deformans, but there is often more synovitis associated. There is rarely the involvement of the joints of the hands as in arthritis deformans. The brunt of the attack in gonorrhœal arthritis often falls especially on one joint, although several joints are attacked. The complement fixation test may be of value.

3. **Gout.**—This may appear as an acute polyarthritis and cause difficulty for a few days, but usually not for long, unless it be one of the cases of gout which has a prolonged course of acute features. The absence of marked changes in the joints and the results of treatment are suggestive. The finding of tophi and sometimes a study of the uric acid excretion aids its recognition. In some cases of chronic gout without any special distinguishing features the diagnosis may be difficult. The relative frequency of the diseases should be kept in mind; arthritis deformans is much the more common.

4. **Some of the Forms of the So-called "Infectious" Arthritis.**—This group may pass over into arthritis deformans, and it is difficult to know where to draw the line. The frequent occurrence of arthritis with or after many acute infections has to be always remembered. It is especially important for prognosis to distinguish them, but this may be difficult until time gives the answer. The knowledge of a source of infection, the results of treating this, the progress of the arthritis, and time are our greatest aids.

Certain forms of arthritis deformans require special mention.

(a) *The So-called Still's Disease.*—This is not likely to give much difficulty, as the picture it presents is usually characteristic in the association of a multiple arthritis, especially with peri-articular changes, marked general glandular enlargement, and an enlarged spleen. In some cases a general tuberculous arthritis may give difficulty, but the x-ray plates and the use of tuberculin should settle the question.

(b) *The "Monarticular" Form.*—This term is only used for clinical convenience. On careful examination slight changes in other joints can usually be found. The greatest difficulty arises in the shoulder-joint where two conditions especially cause error. One is neuritis, arthritis often being regarded as this, and the other is subdeltoid bursitis, which is usually regarded as arthritis. The diagnosis of neuritis may be correct so far as it goes, but it is secondary to arthritis. Careful examination will usually show signs of arthritis. The features of subdeltoid bursitis should be looked for, and it will usually be found that the joint itself is free. In subdeltoid bursitis there is pain when any attempt is made to abduct the arm, or it may be elicited by pressure over the bursa. Occasionally the pain is referred along the bicipital groove or down the arm. There is marked restriction of motion, especially abduction, and internal rotation is usually markedly limited. Sometimes the swollen bursa may be felt. In long-continued cases there may be atrophy of the muscles of the shoulder girdle. Sometimes, however, the exact diagnosis may be difficult, and then the *x-ray* plate is useful.

In the hip-joint there is usually less difficulty, although sometimes the condition is regarded as tuberculous. The tuberculin test is useful. If there be communicated inflammation to the sciatic nerve the diagnosis of sciatica may be made. Knowledge of the possibility of this error is a great aid in avoiding it. Disease of the sacro-iliac joint may cause error. Search for the signs of disease there will usually prevent this.

(c) *Atrophic Form.*—It is often possible to recognize this form before making the *x-ray* examination, which makes the diagnosis certain. The joints show marked degeneration, partial dislocation occurs early, muscular atrophy is often rapid in its appearance and progress, and there may be pronounced grating in the affected joints. In interpreting the *x-ray* plates, it must be remembered that there may be a certain amount of bony atrophy secondary to any continued arthritis.

(d) *Hypertrophic Form.*—While the examination of the joints may serve to distinguish this at a glance, as in the Heberden's nodes, yet in the larger joints the chief dependence must be placed on the *x-ray* plates. In a joint—such as the knee—with marked muscular atrophy above and below, there is often the suggestion that there is enlargement of the bones; the *x-ray* plate frequently shows that this is not the case. Of special forms, the Heberden's nodes rarely give any difficulty, especially if several joints are involved. Injury, however, may have results which closely resemble them, but this rarely affects more than one or two joints.

(e) *Spondylitis.*—This has been discussed on page 928.

There are certain ailments which may cause confusion, these usually being regarded as arthritis deformans. Sprains, pain due to muscular strain, pain due to certain occupations, relaxation of ligaments, flat foot, neuritis, lumbago, and such like, can generally be excluded or recognized by careful examination.

The *x-ray* examination is always of the greatest help not only in deciding as to whether or not arthritis is present and its nature, but also in giving information as to the degree of change in the joint. To estimate this last should always be our aim. To know the form we are dealing

with is important both for prognosis and treatment. To know how much change there has been in the joint is essential for proper and intelligent handling, and should be as much a part of the diagnosis as the determination of the disease itself.

**Prognosis.**—In forecasting the outlook in a disease with so many varying features, it is evident that much has to be left to the judgment of the physician and the conditions in a particular patient. There is no general rule which we can apply. The type of disease makes a great difference as well as the general character of the patient. Speaking generally, the writer is inclined to be more hopeful about the outlook than the majority of those who have discussed the disease. There are certain exceptions which will be noted. Let it be said that there is no intention of minimizing in any way the gravity of chronic arthritis. To fight a winning battle with one of the most intractable of chronic maladies, not as regards life but for the comfort and usefulness of the patient, it is necessary to know the power of the enemy and to have a wholesome respect for the dangers. Perhaps nothing has given more personal encouragement than the results which have been obtained in dispensary practice. To get hold of patients early in the course, teach them what to do, have them work enthusiastically despite discouragements, and finally get through without much damage is cheering. To see a patient grow steadily worse despite all treatment dampens our enthusiasm. Yet it is important both for the physician and patient that the most cheerful view be taken, for both will work harder. The character of the patient must always be taken into account. Some will work hard, others expect everything to be done for them. The condition of the nervous system is often important. High-strung, nervous patients often do badly, and any such condition is usually aggravated by the arthritis.

1. **Peri-articular Form.**—Here there are many factors to be considered. (a) *Early Diagnosis:* This is most essential, in order that proper treatment may be begun early and that no treatment will be carried out which will do harm. (b) *Recognition of the Cause:* If the local focus of infection can be found and properly treated before great arthritic damage is done, the outlook is much more encouraging than when the contrary is the case. (c) *Age:* Generally the older the patient the better the outlook, although this is not always true. In older patients there is sometimes a tendency to slowly progressive degenerative changes. (d) *Station in Life:* While in some cases the possession of wealth and the ability to have expensive treatment, such as massage, is of benefit, yet often the poor, who have the stimulus of necessity, seem to do better. Too often the rich are able to lie back and be waited on, to their ultimate sorrow, while the exertion demanded of the poor may be their greatest aid. (e) *Character of the Patient:* This means a great deal. Those who fight and "never say die," always working, as a rule, do well; but those who lie back and demand that the physician do everything, who are willing to take medicine but not to exercise, generally do badly. (f) *Character of the Attack:* The severity of the initial attack is no guide to the ultimate result. As a rule, in the patients with acute attacks the outlook is much better than in those with a slow progressive process which

advances steadily from joint to joint. The attacks which begin in women about the menopause always demand a very guarded prognosis. The outlook in them is likely to be bad. The acute progressive cases can usually be recognized, and in them a very guarded prognosis should always be given. (*g*) *General Nutrition*: This is important, and in patients whose nutrition is much affected and who do not respond to good hygiene, diet, etc., the outlook is always serious.

There are some local features in the joints which are of aid. (*a*) *Pain*: The degree of this is no indication of the severity of the arthritis, and yet its influence on the general health of the patient cannot be disregarded. Its presence may hinder use of the joints and favor ankylosis. It may be practically impossible for the patients who have severe pains in the feet to walk and obtain sufficient exercise. (*b*) *Changes in the Joints*: With joints in which the changes are entirely peri-articular and cartilage and bone show no change, the outlook is better than when signs of erosion and thinning of the cartilage are evident. Here, however, caution is needful, for the cartilage which shows no change today may do so in six months or a year. (*c*) *Joints Involved*: This is sometimes a help. The elbows if attacked often show marked permanent changes, the wrists and shoulders much less. Marked changes about the knuckles and finger-joints are often suggestive of much future deformity. The hip-joints have a tendency to permanent fixation. The knees easily become flexed, and arthritis of the joints of the feet may be very crippling without much actual change. The neck and jaws usually do well.

In the "monarticular form" the outlook, as a rule, is good for fair, ultimate recovery, but generally one cannot prevent the rather long duration. Some of the patients with marked atrophy of the muscles may be two years before reaching a fair degree of recovery. In the so-called Still's disease, the majority of writers regard the condition as practically hopeless. My experience has been more encouraging, several patients have done extremely well.

In this group certain features are of importance in estimating the outlook. If there be marked muscular atrophy which has come on rapidly, the outlook, as a rule, is more serious. If there be marked early tendency to contracture, it is always well to be cautious. Then the patients with marked general nervous disturbance, as a rule, do not do very well. With this seems to come an intolerance of pain, so that it is impossible to carry out the usual measures to prevent ankylosis and contracture. Naturally in a disease which is so often accompanied by general disturbance of nutrition the condition of digestion is important. In patients who are unable to take enough nourishment or whose digestion is easily upset, one should always give a guarded prognosis.

2. **Atrophic Form.**—In this it may be said in a general way the outlook is always serious. We do not seem to have any means of hindering the advance of the disease, which, as a rule, progresses to more and more disintegration and destruction of the joints. No matter how slight the early changes may seem to be, or how good the patient's general condition, the prognosis should always be guarded. The general condition is often the best guide as to the outlook.

3. **Hypertrophic Form.**—In this the outlook depends on many different features and no general statement can be made. (a) *Heberden's Nodes*: As regards any possibility of absorption of these there is only one answer to give: Once present, they are permanent. However, beyond the appearance and slight discomfort, they rarely give any severe trouble. If they are painful, one can usually assure the patient that this will not last for a great time. Much depends on the patient's avoidance of injury, and this influences the outlook to a considerable extent. (b) *Conditions in the Larger Joints of Elderly Patients*: Although the general tendency is toward slow progression, yet the outlook is in many instances better than one might have expected. It is never safe to be too positive about the outlook in either direction, and it is not unusual to find them doing much better or much worse than one expected. However, as a rule, the possibility of complete recovery is almost out of the question. (c) *Smaller Joints*: Although the condition may not advance very materially and a fairly good prognosis is often justified, it must always be remembered that bony outgrowths having once formed cannot be removed unless by surgical measures, which are sometimes indicated. The hope is that further progress will be prevented, and the patient's condition not become worse than it is. (d) *Spondylitis*: In this the outlook depends very largely on how much of the spine is involved. If the patients are seen in the acute stages a good prognosis can usually be given as regards the freedom from pain, but the amount of disability cannot be estimated. If the process is at all general there may be rigidity of the whole spine, with more or less deformity, although it is always encouraging that the extent of the fixation rarely corresponds with the extent of acute arthritis. In the cases of local involvement, as, for example, in the lower spine, the outlook is generally good if treatment has been begun early. The process is usually confined to a few vertebræ, and the patient recovers with very little disability. The possibility of subsequent attacks involving more vertebræ can never be excluded.

In all forms the general condition is important and the possibilities of good surroundings and proper nutrition must always be considered. As has already been said, the outlook depends a great deal on the patience and perseverance of the patient and physician. The possible effect of some intercurrent acute infection is difficult to forecast. While, as a rule, any such tends to aggravate the arthritis, as, for example, an attack of influenza, yet in some patients the state of the joints is much better after an acute attack of another disease.

In all acute cases two questions arise—the outlook in the immediate attack, and the ultimate result in the joints. As noted, the acute attacks are usually over sooner and often leave but little damage. The general features, such as fever, and the arthritic conditions are both important, but it is never wise to prophesy too much. In the slowly advancing form the prognosis should always be very guarded. As to the ultimate result in the joints, the x-ray examination is always of great value. If this shows peri-articular change only, the outlook is usually good; but if early erosion and destruction of the cartilage is found, one must be very cautious. In cases with marked changes, the question as to possible

restoration of function is also greatly aided by the radiograph. With damaged articular surfaces, displacement, or marked atrophy the outlook is never good. If the joint surfaces are not damaged, even with a good deal of contraction, it may be possible to get back function in a wonderful way. The possibility of aid from surgical intervention should always be taken into account.

**Treatment.**—It is evident that in a disease which has so many different manifestations and methods of progress no scheme of treatment can be laid down which will apply to all patients. Again, it is a disease at times characterized by acute attacks which may end as such or pass over into a chronic stage, or it may be chronic from the onset. Then, too, in its treatment both the general condition of the patient and the state of the joints have to be considered. It is well to emphasize the fact that treatment to be of avail must extend over a long period of time. Dogged perseverance must be shown both by the physician and by the patient. Too often both lose patience and part company, the patient to see if some other practitioner (too often some quack) cannot help him more, the physician perhaps with a feeling of relief that this particular knotty problem has gone to someone else. Perhaps more than of any other disease is it true of this one that the patients rarely go through a very long siege of the malady without seeing many physicians.

The subject has its dark and its bright side. When one sees a patient in whom the disease progressively advances, crippling one joint after another, more or less steadily advancing in many joints despite all treatment, the feeling of hopelessness and helplessness cannot be escaped. At the opposite extreme, when a dispensary patient steadily improves, even under adverse circumstances, one feels that there is some hope for the majority. It may be said that this is no disease to be treated by medicines alone. The physician must be prepared to work hard and keep his patient working hard—often a much more difficult task. The whole mode of life must be supervised and constant attention paid to every detail. There must be much the same attention given to every point as in the case of a patient who has pulmonary tuberculosis.

At the risk of tiresome repetition, the need of early diagnosis must be emphasized again. It seems absurd to insist on this so often did not the history of many patients prove the contrary. The great hope for many of them lies in early recognition and proper handling. Too often the error is made of regarding the disease as "rheumatism," after which almost everything done for the patient is more likely to harm than to help him. To keep the patient indoors, to cut off protein food, to give prolonged hot baths, to administer large doses of salicylates, are injurious as a rule. After the disease has run its course, the effort to restore function to greatly damaged joints must often be fruitless. Early recognition and persistent treatment are all important.

The treatment of a given patient generally has to be considered from two points: (*a*) that of the general condition, and (*b*) that of the joints. On the whole, the former is the more important; the whole is greater than the part, and if the individual is doing badly, his joints are not likely to do well. Every disturbance of the general health should be

regarded seriously, and these patients should take unusual precautions with any slight infection, gastro-intestinal trouble, etc.

*Source of Infection.*—In line with the view that the disease is dependent on some infection, every effort should be made to discover a primary focus, if such exist. This demands very thorough investigation—there is no use in half-way measures. If definite change has occurred in the joints, the removal of the original source of infection will not result in complete recovery, but it will probably stop any advance of the process. The condition of the mouth must be first looked to, carious teeth and areas of infection should receive attention, and pyorrhœa alveolaris be treated. The tonsils should always be carefully examined. If they show obvious infection, there is no doubt as to the advisability of removal. If there be no direct evidence of involvement, the decision is more difficult. Frequently a tonsil which shows no surface evidence of disease, after removal shows deep pockets which contain streptococci. If there is any doubt as to the condition of the tonsils, it is better to give the patient the benefit and advise removal. This should be thorough; nothing short of complete removal by dissection is sufficient. Careful examination should always be made for any focus of infection in the sinuses. Otitis media should be treated if present.

The respiratory tract does not offer many examples of the source of infection being there, yet a persistent bronchitis may have an influence and should be treated. In the same way any gastro-intestinal disturbance should receive attention. The effort should be made to correct any digestive trouble. How much intestinal disturbances may be casual factors we do not know; but, at any rate, for the sake of the general nutrition, if for nothing else, we should try to prevent them. Dysentery especially should be thoroughly treated. Some patients in whom there is no definite intestinal disturbance are helped by free purgation, and others by daily washing out of the colon. Visceroptosis should be corrected by the usual measures.

Next to the mouth and pharynx, perhaps the genito-urinary organs are the most important causes of infection. In males, it is especially necessary to investigate carefully the prostate gland and seminal vesicles. The contents should be massaged out and thoroughly examined. In women, the possibility of any infection of the urinary tract and the condition of the pelvic organs should be determined. Leucorrhœa should be treated. Any source of infection should receive attention.

If local trouble is found anywhere, the measures which are demanded for its treatment should be carried out. The question sometimes arises as to whether any severe local treatment—such as removal of the tonsils—should be carried out while the patient is having acute arthritic symptoms and perhaps fever. If we are correct in attributing the condition to the local focus, there is every reason why this should be treated at once. An analogous case is seen in gonorrhœal arthritis, in which it is equally important to treat the urethritis as the condition in the joint.

*General Considerations.*—This is a debilitating disease, and the importance of this should be kept in mind. No measure should be adopted which can affect the general condition unfavorably or in any way reduce the patient's resistance.

*Hygiene.*—The patients should have all the fresh air and sunlight possible. If able to walk they should be outdoors whenever possible. If unable to do this, perhaps owing to general involvement or acute remissions, they should be kept outdoors as much as can be managed, exactly as the patient with pulmonary tuberculosis. They are often benefited by sleeping outdoors. In cold weather it is necessary to see that they are properly clothed, and the extremities especially should be kept warm. It is important to protect them against sudden changes of temperature. In patients with marked involvement of the feet or hands comfort is often given by having them wear bed socks or woolen gloves at night. Some patients are helped by periods of rest each day.

*Diet.*—In general, it may be said that as nourishing a diet as possible should be given. Individual peculiarities and special conditions have to be considered—that is common-sense. In some patients who have been heavy eaters it is well to diminish the amount. The diet should be chosen which agrees best with the patient. In addition to the regular meals, it is well for a time to give extra nourishment between meals and at bedtime. This may be in the form of milk, eggs, cocoa, etc. One very common error is to diminish or cut off as far as possible all the proteins from the diet, it being often suggested that the disease has something to do with uric acid. This almost always results in harm, and sometimes very serious harm. The writer would lay special stress on this, and emphasize the injury done by restricting the diet for no other reason than this. If anything is to be cut off from the diet it is usually better to reduce the carbohydrates, as many of the patients are subject to some digestive disturbances which are aggravated by too much of this form of food. A full protein diet is usually indicated, and, as a rule, the fats are well taken. Some are helped by the use of the fermented milk preparations. If the patients are accustomed to alcohol there is no reason why small amounts should not be given. As a rule, large amounts of water should be taken to favor elimination. There is no special virtue in any particular water, except that some patients can be induced to drink more when some name is attached.

*Bathing.*—Apart from some of the measures of local hydrotherapy, such as douches, compresses, etc., bathing is not of value, as a rule, in the *acute* stages; in fact, it is often positively harmful. Prolonged hot baths are often debilitating and should not be given. A sojourn at various springs is usually of more value from the change than from any special virtue in the baths.

*Massage.*—The majority of patients are helped by general massage; that of the joints will be discussed later. It aids the general condition and helps to lessen the amount of muscle atrophy. Care should always be taken to see that it is not given too vigorously, a mistake often made. Many of these patients are exhausted after any but the lightest rubbing, so that this, certainly at first, should be as light as possible, and increase in the vigor with which the massage is given must be very gradual. Two or three times a week are usually enough for general massage. The use of some form of vibratory movements is useful in some cases.



*Electricity.*—This is sometimes used, and some favorable reports have been given, especially from the use of electric light baths, which probably owe their effect to their influence on nutrition.

*Radium* may be useful in some cases but the results thus far are not very encouraging.

*Climate.*—There is no doubt of the benefit of a change of climate, especially from the North to the South in winter. Whether this is due to anything more than the possibility of being more outdoors and leading a life in the open air is difficult to say. Certain patients are more apt to have recurrences in cold weather, and improve immediately with a change to a warmer climate. As a rule, a dry, equable climate suits the patients best.

*Vaccines.*—If we are correct in supposing that an infectious process of some kind is the important etiological factor and the organism can be obtained, the use of an autogenous vaccine may be helpful. Stock vaccines and serums are not of much value.

*Medicinal.*—That we possess any drug which essentially influences the disease there is no evidence. Naturally, in a debilitating disease, it is important to keep the blood in as good condition as possible, and for this iron and arsenic are perhaps best, the iron being given as Bland's pills and the arsenic as arsenous acid combined with Bland's pill or in the form of Fowler's solution. Many patients do very well on the syrup of the iodide of iron, which may be given in full doses over a considerable time. Some few patients are benefited by potassium iodide given persistently. That any of the salicylate preparations are of value is doubtful. They may be used for pain, but their long-continued use is not advisable. Guaiacol carbonate in doses of gr. v (gm. 0.3) is strongly advised by many of the English writers. Some have obtained good results from iodine in the form of the tincture in doses of m℥ to i℥, especially in acute attacks. Thyroid, thymus and pituitary extracts given persistently seems to have helped some patients.

Pain often requires general treatment. Few patients escape this entirely, but in many the various local measures are sufficient to give relief. Some patients have such severe general pains that drugs must be given. The salicylates, acetylsalicylic acid in doses of gr. viij (gm. 0.5) or more, guaiacol carbonate, gr. v (gm. 0.3), antipyrine, gr. iij (gm. 0.2), sometimes combined with codeia, gr.  $\frac{1}{4}$  (gm. 0.016), may be tried. Morphine should not be given, as there is danger of forming a habit.

Marshall has drawn attention to the effect of ether, administered as a general anesthetic, on the arthritis especially when very acute. The ether is given in the usual way for about fifteen minutes. The pain is sometimes markedly diminished and the condition of the joints improves. The period of improvement varies from a few days to two weeks. This is worthy of a trial in severe attacks.

*Treatment of the Joints.*—Here only general indications can be given and every patient must be carefully studied. It is always important to be sure that we are not doing any harm. The various forms demand different handling, and a degree of exercise which is of great help to a patient with marked peri-articular changes may be harmful to one

with hypertrophic changes predominating. As a guide to treatment, the study of the *x*-ray plates is of great aid. Thus, if there is marked destruction of the cartilage or hypertrophic bony changes it is evident in a general way that rest is important and exercise harmful, which if the changes are peri-articular we are usually safe in advising use.

**Peri-articular Form.**—In general terms it may be said that exercise and use of the joints are beneficial. This is probably for several reasons, the nutrition of the structures is better, the risk of ankylosis and muscular atrophy is diminished, and the chance of contractures lessened. This is well seen in the temporomaxillary joint, which is relatively often affected, but which rarely shows ankylosis. Talking and chewing are two exercises which keep it from fixation. One caution is important—use should not be begun too soon. Rest should be the rule so long as the condition is acute. Yet even here there are exceptions, and sometimes it is well to begin gentle massage and movements before the acute features have entirely disappeared. Care should be exercised in the use of fixation as a therapeutic measure. Atrophy, adhesions, and contractures may appear with great rapidity.

**Passive Movements.**—These may often be begun early. They should be carried out gently at first and increased as the condition warrants. The production of pain is not necessarily a contra-indication, and especially when there is a tendency to contracture. Some patients have pain at first on the gentlest movement, but this usually lessens. The early use of passive motion is of great value in preventing adhesions.

**Massage.**—This is often most helpful, but must be begun gradually and increased slowly. It is important to oversee this point, as many of those giving massage are likely to be too energetic. Gentle kneading and circular movements about the joint are the most useful. For the muscles above or below the joint more vigorous measures may be used. If there is severe pain it is often advisable to have the rubbing done after the use of measures, such as hyperemia, which ease the pain. The patients can often be taught to do a certain amount themselves. They can work on the hands and wrists very well after being shown how to perform a kneading motion with the thumb on one surface of the hand and the fingers on the other. One advantage of this is that they are doing good to both hands at the same time. They can sometimes work at the elbows, knees, or feet in the same way.

**Exercises.**—These are useful. The patient should be started at very simple ones in the beginning. Thus, with the hand they may begin by making a fist and alternately spreading the fingers and bringing them together. A rubber ball with a small hole in it which they can squeeze in the hand is of help to many. Various movements at the wrists, elbows, and shoulders may be done. Lifting the shoulders, rotating the arm, raising it up, etc., may all be tried. In the same way the knees, ankles, and toes may be exercised. It is sometimes an advantage to have them keep a record of the exercises and the number of times each was done. Certain forms of mechanical apparatus may be used in the giving of exercises. In all of these it is important to go slowly at first and always stop short of fatigue. The patient's strength should be estimated, and

the number of exercises and the number of times each is to be done carefully ordered. Increase should be very gradual. One great advantage of the exercises done by the patient is that it gives them a feeling of doing something for themselves and of making a personal fight.

*Counterirritation.*—This is often of great help, both for the pain and to aid the local nutrition. The Paquelin cautery lightly applied, blisters, mustard, or iodine may all be used. Repeated small blisters are usually better than one large one. Baking may be considered here. This may aid the blood supply, but it is chiefly of help in easing pain. Care should be taken not to give the treatment for too long a time; twenty to thirty minutes is usually enough. The temperature in the oven may be raised to 350° F., but there is usually no advantage in reaching the highest temperature which the patient can stand. It is well not to cover the joint with too much material. Care should be taken not to allow the patient to become chilled afterward, especially if there has been sweating. The frequency with which baking is done depends on the severity of the condition and how the patient reacts. Once a day is generally sufficient for severe cases; in others two or three times a week is sufficient.

*Hyperemia.*—This is sometimes of great aid both in influencing the process in the joint and relieving pain. No rule can be made as to whether active or passive hyperemia will afford the greatest relief in a given case. In using the Bier method the bandage should be put on above the joint with sufficient pressure to interfere with the venous return, but not with the arterial flow. The necessary pressure can usually soon be determined for each patient. At first the bandage is left on for an hour at a time, but this can be increased rapidly until it is on for eight, twelve, or even twenty hours. Some patients prefer to have it put on at bedtime and left on during the night. It is not uncommon to have some transient œdema as a result, but this need not cause any uneasiness. Another method is to empty the limb of blood as much as possible by elevation and stroking toward the body, after which a bandage is applied, with pressure sufficient to stop the arterial flow. This may be left on from thirty seconds to one minute, and is then removed. This should not be done more frequently than once a day. The use of the suction methods for producing hyperemia is not of much value.

*Prevention of Deformity.*—This is most important, and prevention is much easier to carry out than cure after deformity has occurred. Use of the joints and local exercise and massage are perhaps the greatest aids. Every effort should be made to prevent contractures, which are usually most apt to occur in the elbow and knee. By perseverance much can be done, but in some patients it seems almost impossible to prevent contractures, owing to the pain caused by any manipulation or the marked constant contraction of the muscles. In such it may be well to put the joint in a splint for part of the day or use light extension for some hours each day. The danger of putting joints up in plaster casts for several weeks should be recognized, and this is rarely advisable. In some patients the injection of oil into the cavity of the knee-joint may be of value. Persistent exercises and passive motion are helpful for the shoulders.

For the hands a certain amount can be done by the use of light splints, well padded, which may be worn at night. The tendency to flexion at the first interphalangeal joint and to hyperextension at the proximal joint can be considerably lessened by this. In some patients it seems that the tendency to ulnar deflection can be diminished by the use of a light splint, although one must not hope for too much from this.

**Treatment of Deformity When Present.**—At first sight the hope of doing much for already established deformity often seems slight, and yet it is surprising what can be done by persistent effort. The employment of various forms of machines for giving the exercises is of great service in these cases. The patients should be encouraged to work themselves in every possible way. Sometimes putting them in a rocking-chair and letting them rock gently, for gradually increasing periods, is of help for contracture of the muscles of the leg. Massage may be steadily used. Extension is sometimes useful for flexion of the knees, and gentle forcible straightening of the fingers, with the use of splints, often helps deformity of the hands. In some cases surgical measures are of value. In deciding as to these it is important to study the x-ray plates and determine how much damage has been done to the articular surfaces. Contracted tendons may be divided and the limb straightened. If this is done the joint should not be left in a plaster cast for some time, but passive movements should be begun early. In the knee-joint the injection of oil into the cavity is sometimes of assistance in aiding an increase of movement. Excision of a joint may be advisable, as when one knee is markedly contracted and the other in fair condition. It is well to try and have the patients go about as much as possible—at first with assistance and crutches, then with the crutches themselves, and later with one crutch and a cane.

**Villous Arthritis.**—This may demand special treatment by bandages or apparatus, but if these do not aid it may be well to advise surgical measures. As a rule, removal of the hypertrophied fringes gives great relief and improved function of the joint.

**Atrophic Form.**—So far as regards treatment this is the most unsatisfactory form, as it is too often progressive despite all treatment. Every effort should be made to keep up the general health by diet, hygiene, etc. With the destruction of the cartilages and marked thinning of the bones, use should be somewhat restricted for a time, but some of them seem to be better with moderate use. Every patient must be carefully studied and while it is difficult to lay down any definite rules, generally if there is any doubt it is better to incline to rest. Hyperemia, very gentle massage, and baking help the patient's comfort, and should be tried. Very often a change to a dry, equable climate gives good results.

**Hypertrophic Form.**—In this the matter of rest is important during the acute stages, when, as a rule, exercise is harmful and rest is useful. The affected joint is usually helped by some protection. Sometimes one has to take a middle course, as in the elderly patients with hypertrophic changes especially in the hip-joint. If they walk too much, there is danger of stirring up the process and making it more acute; if they keep too much at rest, there may be danger of fixation. Going up and down

stairs is often especially harmful to them. With overgrowth of bone it is evident that harm may be done by active or passive motion. Rest may often be secured by mechanical support, which may be useful even when it does not secure complete fixation, although the latter is usually advisable in very acute conditions.

(a) *Heberden's Nodes*.—These sometimes require treatment, although nothing can be done to prevent their appearance. Increase in size is often due to trauma or irritation by use, rubbing, etc., so that the patients should be especially warned against injury, and if acute features are present, such as pain and redness, they should be carefully guarded against any trauma and kept as undisturbed as possible. If there be pain and tenderness, the most efficient treatment is usually hydrotherapy in the form of wet compresses covered with some form of protective, put on at night and left until morning. The patients should be cautioned not to rub the joints, and to limit their movements.

(b) *Spondylitis with Hypertrophic Changes*.—Rest is usually the greatest aid in treatment. To secure this, a plaster jacket or some form of apparatus should be worn. Not infrequently this causes increased pain for a few days, and may have to be changed, but relief usually comes in a short time. The jacket should come up to the shoulders and well down over the hips. Some patients are relieved by a form of corset, carefully adjusted, or by a belt made of webbing. These are generally advisable after the acute stage is over. Caution should always be given as regards the danger of trauma or strain. Lifting of weights or severe exertion, especially in strained positions, in fact anything which may throw a sudden strain on the spine and its muscles, should be carefully avoided. In the patients who suffer especially when in bed, care in the kind of mattress, adjustment of small pillows, etc., may be of use. In the patients who have a general process which is recognized early, an attempt may be made to have the spine in the most favorable position before fixation occurs.

In the more generalized forms it is well to save the joints during the acute stages. This is not necessarily always complete fixation, but certainly excessive or even normal use should be avoided. After the acute features are over, such a joint as the knee is often helped by some support, such as an elastic covering. If bony exostoses are causing mechanical difficulty, surgical measures may give some relief; bony spurs, or the osteophytes about the joint, may be removed.

In all forms attention should be paid to associated conditions, such as flat foot, knock-knee, etc. The use of weights may relieve deformities greatly. The orthopedic surgeons have done much to aid these patients.

In conclusion, emphasis should be laid on the need of perseverance and patience in treating patients with these chronic joint conditions. Early recognition, persistent and intelligent fighting, and hard work often accomplish wonders.

### FIBROSITIS.

Under the term myalgia or the unsuitable one of "muscular rheumatism" are grouped a variety of conditions but little understood and

which have perhaps in common only the symptom of pain. These are comparatively frequent in occurrence, vary greatly in severity and acuteness, and for the present are often difficult of explanation. The acute forms we see especially as torticollis or lumbago. The more chronic forms are associated with more or less pain and at times with varying degrees of disability and stiffness. The essential nature of the condition is in doubt. By many it is regarded as associated with rheumatic fever, or being a manifestation of the rheumatic state. Others regard it more in the nature of a neuralgia, some as connected with a diathesis or gout. The changes are apparently very largely in the white fibrous tissue, consequently the tendons, muscle and nerve sheaths, periosteum, the various fascia, and the fibrous portions of the ligaments are involved. Consequently, for many of this group the designation fibrositis is suitable.

The *etiology* probably differs in the acute and chronic forms. The acute form occurs more often in men and in adult age. It may follow exposure to cold, but here, as in other forms, trauma often plays a part. Thus, in the production of torticollis, if a patient is lying with the head in a bent position the condition is apparently more likely to develop. It is, however, especially in the production of lumbago that trauma seems to be important. Some strain, as in lifting or working in a bent position, is a common contributing factor. This, with exposure, is a common cause in laboring men. The more chronic forms are commoner later on in life and occupations involving exposure are apparently important factors. In both the acute and chronic forms one attack seems to predispose to another. As a result of the thickening of the muscle sheaths a considerable degree of disability may be produced, following which there may be muscular atrophy and a certain amount of contraction.

The essential *pathological* lesions are apparently in the white fibrous tissue, and the condition is probably of an inflammatory nature. There is a serous exudation in the affected parts, which later may be followed by proliferation of the fibrous tissue. This may be absorbed entirely, but if long continued will be more or less permanent and may extend over a considerable area. As a result of these, various nodules have been described, the histological character of which has been described by Stockman. Thus, in one patient a small nodular swelling in the buttock was excised, and found to consist of a portion of the perimysium of the gluteus medius muscle. Sections showed it to consist of the fibrous sheath of the muscle hypertrophied and oedematous. In place of the fibre, tissue penetrated between the muscle fibres, some of which were degenerated. The small vessels showed periarteritis and endarteritis. There was no evidence of any very acute inflammatory reaction and no organisms were found in the tissues.

It is probable that a number of factors enter into the production of this condition. The changes described by Stockman suggest either chronic low-grade infection or the influence of toxins. Many cases are probably due to absorption from a focus of infection.

As to the *symptoms*, in the *acute* form the main features are pain and disability often coming on suddenly. The pain may be sharp and fairly

constant or dull, and becoming severe on movement, or when the muscles are placed in certain positions. Special features depend on the locality affected and certain forms are fairly well recognized.

1. *Torticollis* affects the muscles of the neck laterally or posteriorly. The pain is generally very severe, and the patient holds the head to one side. The condition is usually unilateral. It may persist for a few hours only or for several days.

2. *Lumbago* affects the muscles of the back. The onset is usually sudden, and often after some muscular effort which involved bending or lifting. The pain may only be present on movement, but is sometimes so severe as to incapacitate the patient and prevent any movement. The duration is very variable, but is usually a few days. Some individuals have regularly recurring attacks. As with torticollis, a striking feature is the frequent prompt response—although the effect may be only temporary—to local counter-irritation.

3. *Pleurodynia* involves the intercostal muscles and sometimes the pectoralis and serratus muscles. The pain is severe and greatly aggravated by any movements. The respiratory movements are restricted on the affected side. Pressure may cause severe pain.

4. Various other forms are described as *scapulodynia*, *dorsodynia*, etc. Their general features are much the same. A certain set of muscles may apparently show a tendency to repeated attacks.

Constitutional symptoms are not common. Fever is rare and the pulse rate is not increased.

The *chronic* forms are distinguished especially by aching, soreness, or pain, and varying disability which consists chiefly in stiffness and inability to perform certain movements. These may be more or less constant, or come and go for various periods. In severe cases they are present constantly, and exacerbations occur, as, for instance, after exposure. Sudden muscular movements are especially apt to cause pain. In addition there may be severe pains which are apparently neuralgic in character.

The muscles may be tender on pressure, and in some cases definite areas of induration may be felt. Stockman lays considerable stress on these, and describes some as being soft and ill-defined, while others are firmer. They are usually tender on pressure. He describes the commonest seats as the lumbar aponeurosis, the calves, fascia lata, trapezia, insertion of the deltoid, the biceps, the intercostal and pectoral muscles, the glutei and soles of the feet. With time, considerable disability and marked loss of function may result.

The recognition of this condition, as a rule, does not offer much difficulty, with the exception of lumbago. Here the possibility of arthritis of the spine, sacro-iliac-joint diseases, tuberculous disease of the spine, or secondary malignant growth in the spine must be kept in mind. Careful examination with the possibility of these kept in view and the need of excluding them before a diagnosis of lumbago is made, are the most important points.

Concerning *treatment*, as regards the general management, it is usually well to open the bowels thoroughly and to see that the patients take large

amounts of water. For the pain some of the salicylate preparations are generally most efficacious. Sodium salicylate, in doses of gr. x to xv (gm. 0.6 to 1), or acetylsalicylic acid, in gr. x (gm. 0.6) doses, may be given. For some patients small doses of colchicum are most useful. Guaiacum and potassium iodide give considerable relief in some instances. As to local measures, the most important is rest to the affected muscles. This may be secured by mechanical means, especially strapping. It is probable that the often used plaster, especially to the back, acts in this way as much as any other. Local counterirritation and the application of *heat* are particularly helpful. The Paquelin cautery may be applied or blisters used. Acupuncture is very helpful in lumbago; either sterilized needles or hatpins are inserted in the lumbar muscles, after cleaning the skin, and left there for five minutes. In some cases the use of the constant current is very helpful. In those subject to repeated attacks, every care should be taken to avoid the exciting cause, which in many patients is exposure to cold. Some of those who are susceptible find that a Turkish bath taken at the very onset frequently has a marked effect in limiting the progress.



## CHAPTER XXVI.

### OSTEOMALACIA.

By GEORGE DOCK, M.D.

**Synonyms.**—*Mollities ossium*; *fragilitas ossium*; *ostitis malacissans*; *malacosteon*; *halisteresis ossium*; *Osteomalacie* (German); *ramolissement des os*, *ostéomalacie* (French); *rammolimento delle osse* (Italian).

**Definition.**—A chronic disease characterized by pain and muscular weakness, and by decalcification and absorption of bones already formed, with bending or fracture, and consequent deformity.

Osteomalacia has been considered rare everywhere, but with the more accurate recognition of its symptoms this has been changed, although it is too early to speak positively of the existence or absence of the disease, which should be more carefully looked for in all parts of the world. It will probably be found that mild cases occur in many places, but the existence of certain severe foci will probably not be less striking than it now appears.

**Etiology.**—The explanation of the endemic occurrence of osteomalacia was naturally sought in the soil and water, but has been abandoned.

Unfavorable hygienic surroundings in the dwelling house or work place, dampness, insufficient clothing, trauma, psychic shocks, and poor food, as sour rye, have all been looked upon as causes, but they obviously can be concerned only in an indirect way. The majority of patients belong to the poorer classes, but there are exceptions.

**Sex.**—Women show a remarkable predisposition to osteomalacia. Among older statistics a proportion of ten to one was general. Latzko more recently (1897) found 5 men among 120 cases. The difference cannot be explained by the influence of pregnancy, as women who have not borne children are also more disposed to the disease than men.

A relation between osteomalacia and pregnancy, the puerperal state and lactation can be seen not only in the human species, but also among lower animals (cattle, sheep). Multipara and especially those with rapidly following pregnancies are more disposed than others. The tendency to dental caries, to slow callus formation after fracture, and the craving for earthy substances, so often noted in pregnancy, may be looked upon as having some relation to the greater incidence of osteomalacia. The belief that osteomalacia occurs chiefly in women of unusual fertility is based upon a statistical error. Heredity plays no part; a family tendency has rarely been observed. The true relation of osteomalacia to rickets is not known. They occur together in certain localities, but considering the frequency of rickets this is not remarkable.

**Age.**—The majority of cases begin between the twentieth and thirtieth years. Cases before twenty are rare, but Drake reports the case of a

Brahmin, married at eleven, pregnant at fourteen, and with severe osteomalacia at eighteen. The observations of von Recklinghausen, Rehn, and Juergens make it probable that osteomalacia can occur in childhood, distinct from rickets, or even *in utero*. It may be combined with rickets. Ziegler denied the non-rachitic nature of the changes in one of von Recklinghausen's cases. Juvenile and virile rickets sometimes follow trauma and infectious diseases. Langendorff and Mommsen<sup>1</sup> report an interesting case. Senile osteomalacia is rare. Pierart, out of 247 cases, found only 22 above the age of forty-five. The symptoms may be misleading and diagnosis impossible, as in virile osteomalacia. In the case of Davis,<sup>2</sup> a man, aged thirty-three years, had complained of weakness and pain in the upper extremities for a year, and fractured the left arm by a fall. There was no union. Exploration led to a diagnosis of sarcoma by examination of tissue from the seat of fracture. Careful differential diagnosis seemed to confirm this, and the arm was removed at the shoulder. Skiagrams had been made, but were unsatisfactory. The later examination showed the process was osteomalacia.

**Pathology.**—There is no unanimity regarding the pathology of osteomalacia, and many facts point to a variety of causes and modes.

An important suggestion grows out of the observation of Hanau on "physiological osteomalacia," a porosity of bone not infrequently found, especially in the pelvic bones, in pregnant women. From this it would seem an easy step to the pathological osteomalacia of pregnancy and lactation, and especially when we consider the need on the part of the growing infant for lime and phosphorus and its supply in the milk. The hyperemia of the bones of the pelvis during pregnancy, the changes that occur in the bone marrow, the remissions of the disease during menstruation and relapses in new pregnancies, all go far to indicate a close relation of osteomalacia to the genital functions of the female. Fehling's brilliant hypothesis, that the disease is due to a morbid activity of the ovaries, leading to passive hyperemia and absorption of lime, has in so many cases been confirmed by the effects of oöphorectomy, that it must be considered as partly, at least, applicable. On the other hand, it is negatived by many cases of failure to recover after operation, by relapses, and by improvement following Cæsarean section, or even anesthesia. Latzko saw chloroform anesthesia followed by temporary improvement in ten cases. On the other hand, improvement has followed in cases of oöphorectomy under ether and under local anesthesia. Cases of infantile and senile osteomalacia, as well as cases in men, are not to be explained on the ovarian theory, which, of course, still leaves to be explained the remarkable geographical conditions.

The nervous-metabolic theory of Pommer, though somewhat strengthened by more recent observations on the trophic disturbances of bones, and the theory of Comby, of nutritive disturbances of bone by fermentation products from a dilated stomach, cannot be generally applied.

E. Hoennicke<sup>3</sup> has advanced a thyroid theory, based on many facts relating to the thyroid gland and its functions. He saw a case of osteo-

<sup>1</sup> *Virchows Archiv*, 1877, Band lxix.

<sup>2</sup> *Annals of Surgery*, 1904, xi, 225.

<sup>3</sup> *Ueber das Wesen der Osteomalacie*, Halle, 1905.

malacia in a man, aged forty-seven years, who also had exophthalmic goitre. Similar cases have been reported by others. Hoennicke shows a close relation geographically between osteomalacia and goitre; he found an association of goitre in a considerable number of cases of osteomalacia, and he points out the well-known trophic and metabolic relations of the thyroid to the bones, and the mutual relations of the thyroid and ovaries.

Erdheim<sup>1</sup> calls attention to the relation between lime metabolism and the parathyroid bodies and its possible bearing on osteomalacia. On examining the parathyroids from eight cases of puerperal osteomalacia he found evidences of hyperplasia, followed by atrophy and small-cell infiltration. The occurrence of tetany in osteomalacia adds interest to the observation, and, as Erdheim points out, suggests that the parathyroid hyperplasia in osteomalacia may be due to an increased demand upon the glandular function on account of toxic substances produced in the ovaries or some other organ.

Cristofoletti suggests that the morbid ovarian function causes adrenal hypofunction. Falta suggests there may be absence of the physiological hyperplasia of the anterior lobe of the pituitary body. In osteomalacia adrenalin does not produce glycosuria as readily, or act upon the heart and circulation as in health.

Infection has been asserted to be the cause by many authors, and seems highly probable, but positive evidence is not yet available. The occurrence in the previous histories of severe infectious diseases, and the frequent relation of such infections to the bone marrow, are suggestive.

Laufer,<sup>2</sup> in a valuable study of the subject, suggests the following as a working theory: Osteomalacia can be looked upon as a degenerative disturbance of nutrition in the bones, in the general sense that all degenerative nutritional disturbances are characterized frequently by diminution or disappearance of tissue and by diminution of function. Etiologically, it may be supposed that various obscure factors—improper or insufficient food, trauma, repeated pregnancies, etc.—cause retrogressive changes in the bones. The changes in the bones, decalcification and new formation, are very characteristic of bone physiology, as seen after fractures, and are at first reparable, but continued action of the noxa or other unfavorable conditions permits the characteristic changes of osteomalacia, that may be looked upon as a dystrophy peculiar to the osseous system. The possibility of recovery under various conditions leads us to suppose the dystrophy is independent of irreparable lesions of bones.

The *bones* in osteomalacia, as the name signifies, are soft. At autopsy they can be squeezed or twisted, sometimes feel like wet paper, and cut without resistance. They look and feel greasy. The difference between the brittle and yielding ("waxy") varieties is unimportant. On section the bone tissue proper is much reduced, both in the compact and spongy part. The bones are often altered in shape, generally or in part. There are often cystic cavities of various sizes in the bones, containing clear or yellow or red serous fluid, or dark masses of more or less altered blood.

<sup>1</sup> *Ueber Epithelkörperbefunde bei Osteomalacie, Sitzungsbericht der kais. Akad. der Wissensch. in Wien, Math.-naturw. Klasse, 1907, Band cxvi, Abt. III.*

<sup>2</sup> *Centralblatt für die Grenzgebiete der Med. und Chir., 1900.*

The marrow is yellow and fatty for the most part, sometimes hyperemic or lymphoid in appearance. In some places only the periosteum preserves the original shape of the bone. The periosteum is often thick, hyperemic, and in stripping shows the surface of the bone rough, the Haversian canals sometimes wide and filled with serous fluid. In consequence of the loss of bone tissue the specific gravity is much reduced, from 1.877 to as low as 0.721. The bones permit the passage of Röntgen rays with unusual ease, depending on their decreased density. The process of absorption begins in the marrow cavity and extends outward.

Microscopically, besides the increased sponginess or actual loss of bone tissue, there is often a border of osteoid tissue along the bone. From the fact that this stains red with carmine it has been called the "carmine border." This is recognized as decalcified bone, as von Recklinghausen set forth. The margin of normal bone is not even or systematic, but very often forms a zigzag or irregular line. The decalcified parts at first contain canaliculi and lamellæ, and show at the margins between the calcified and non-calcified parts characteristic lattice-like figures, due to the presence of air in the canaliculi and the spaces occupied by the bone corpuscles and their processes. Hanau's view, that the lattice figures may correspond to defective calcification has not been verified by others. The decalcified parts become softened, sometimes fibrous, like asbestos, sometimes absorbed, leaving spaces filled with marrow, or with mucoid or granular material. Newly formed bone tissue is deposited in various parts, but in general in small amounts. The marrow is fatty or lymphoid, and with giant cells and osteoblasts in varying proportions. Hemorrhages and pigmentation from old hemorrhages cause a great variety of color and of microscopic structure. In some areas the marrow is gelatinous or even watery. The softening, and especially the irregular and large cystic areas of decalcification, explain the more striking deformities.

The bones most affected are those of the pelvis. The sacrum is pushed forward and downward, the posterior parts of the iliac bones with it, while the acetabula are pushed up and inward by the heads of the femurs, the symphysis being squeezed forward like a beak. Other deformities in the shape and position of the pelvis depend upon the position in which the patient remains. The most extreme deformity gives the pelvic cavity the shape of a clover leaf or a Y. In patients who remain long on their backs the pelvis is flattened from before backward. The narrowing does not always act as an obstruction to delivery, as the bones are sometimes elastic ("rubber-pelvis"). Hugenberger thinks that such cases occur in 30 per cent. of osteomalacia. Casati, out of 42 cases, was obliged to do Cæsarean section only twice, and Fehling advised delay in operating for similar reasons.

*Vertebræ.*—The normal curves are exaggerated, especially in the cervical part, so that the chin may rest upon the sternum. Lateral curvatures are frequent, but vary with the habitual position of the patient. Pregnancy often causes the most pronounced deformities. The whole vertebral column is shortened, especially in the lumbar portion. The body sometimes shows a constriction and duplication at the waist, as if the parts had been telescoped.

The ribs are often broken from muscular action, in breathing, or from pressure. They may in part overlies each other. The sternum is often curved or bent. The thorax in general is usually compressed in various directions, causing in time displacements of the heart and lungs, with dyspnoea, asthmatic attacks, and palpitation.

The long bones of the extremities are affected late in most cases, but may be early, causing spontaneous fractures, with sometimes large but usually imperfect callus, or leading to coxa vara, genu valgum, or flat foot. The cranium is rarely affected.

Litzmann has made the following table of the relative frequency of the affection of various bones:

	Puerperal.	Non-puerperal.
Pelvis . . . . .	82	40
Vertebrae . . . . .	46	40
Thorax . . . . .	26	37
Lower extremities . . . . .	15	36
Upper extremities . . . . .	10	30
Head . . . . .	7	24

The joints are not affected. The muscles are atrophied and degenerated, sometimes to a remarkable degree. The nerves of the extremities were also found degenerated by Schlesinger.

The ovaries show no constant change. Fehling, who first attracted attention to the ovaries, described hyperemia, but Winckel and others attributed that to accompanying malpositions. Fehling described hyaline degeneration of stroma, follicles, and bloodvessels, but without proving any of the changes characteristic of or peculiar to osteomalacia.

**Symptoms.**—The beginning of osteomalacia is usually impossible to recognize. Sometimes the increase of symptoms during pregnancy or the puerperal period permits a relatively easy diagnosis, but in most cases the disease is far advanced before any suspicion of its existence is entertained. The rarity of the disease has much to do with the present condition of diagnosis, and a knowledge of the early symptoms and a keener search would result in the more frequent discovery of mild cases.

The earliest symptom is pain. This is usually called rheumatic, but varies in character from dull to neuralgic. In pregnant women it is often felt in the pelvis, especially the sacral region; in others in the thorax, back, or one or more extremities. Movement, but also remaining quiet for a long time, is likely to increase it. The pain is often spontaneous, sometimes nocturnal, but can usually be provoked by pressure on certain bones, especially the pelvis and vertebrae. Tenderness over nerve trunks is sometimes present. Girdle pain has been noted. With the pain there is usually a feeling of weakness and often a subjective and objective stiffness, with contracture or spasm of certain muscles, as the iliopsoas (Renz and Koppen) or the adductors of the thighs (Latzko). From various causes the gait becomes waddling and uncertain; the body sometimes leans forward. Sometimes the gait is spastic, and then the patellar reflexes are usually increased, and there may be intention tremor. If at the same time there is paresthesia, an erroneous diagnosis can easily be made.

If the patient is examined at this time, naked, as is necessary in order to avoid error in all suspected diseases of the motor apparatus, it may be found that besides confirming the statement of the patient or friends that she is shorter, some distinct deformity can be detected. There may be lordosis, or other curvature of the vertebræ, or change in the shape of the thorax, or a furrow in the lumbar region, or a change in the position of the hips and the shape of the symphysis pubis. The linea innominata may be straightened, the diameters of the pelvis altered, but with the conjugate very little or not at all lessened. Examination of the muscles often reveals tremor, fibrillary or gross, or spasm or contracture. Paresthesia in some cases, especially in the legs, and with other nervous symptoms may be due to alterations in nerve trunks from the deformity of the spine and pelvis.

The digestive functions in the early stages are usually not affected. Menstruation and conception are often normal. The difficulties of confinement need not be described here, but it may be pointed out that the fear of conception is not now so great as it has been in the case of osteomalacia patients.

The blood shows no characteristic changes. It becomes chloranemic, with reduction of both coloring matter and number of red cells. The eosinophile cells are sometimes slightly increased, sometimes diminished or absent. Lymphocytes vary; sometimes the large forms are increased. Myelocytes sometimes occur. An increase of the resistance of the red cells has been asserted. The alkalinity of the blood is not always diminished; it may even be increased.

The urine often contains excessive amounts of calcium and phosphates, but these vary without distinct relation to the clinical features. Lactic acid is rarely present, and not characteristic. Bence-Jones's albumose was first found in a case of (probable) myeloma, erroneously called osteomalacia. Since then it has often been found in cases of myeloma. In Dock's case it was found, without other explanation than the osteomalacia.

The general and proteid *metabolism* in osteomalacia is not altered except as the result of associated conditions. The bones become poorer in calcium and other mineral constituents, especially calcium phosphate, and richer in organic matter, especially fat. The earlier statements regarding lactic acid in the bones are now known to be erroneous. The lime leaves the body through the kidneys and intestines. Phosphate calculi are important factors in the urinary excretion. Excretion and retention of calcium and phosphates vary, however, even without treatment, and no conclusion regarding the process in the bones can be drawn from the condition of the urine.

In the further course of the disease, deformities of all the affected bones become most pronounced, and the body assumes the most bizarre attitudes. The skin becomes sallow and flabby. The muscles waste, fractures and callus may add to the deformity. Œdema, dyspnœa, palpitation of the heart, fever, and sweats may be present in various degrees and combinations. Bronchitis and bedsores indicate the extreme cachexia. Dysuria is a frequent symptom. Renal calculi may be passed in large numbers and cause intense agony.

There are many *varieties* of osteomalacia. In rare cases it runs a rapid course, a year or less. In most the course extends over five, ten, or more years, and in this time remissions may occur, lasting many months or years, or recovery may ensue at any time under treatment, rarely spontaneously. Although pregnancy and even menstruation usually make the symptoms worse, improvement or even recovery has been observed in the former. Death is usually due to exhaustion, cardiac weakness, bronchopneumonia, or some other infection. Bedsores often influence the exhaustion and infection. Exophthalmic goitre and myxœdema have been seen with osteomalacia.

**Diagnosis.**—In the early stages the differential diagnosis cannot often be positive, even in an endemic focus. The rheumatic or hemorrhoidal pains of pregnancy must be excluded by careful examination of their seat and course, and the presence or absence of bone tenderness. Later, lateral sclerosis, transverse myelitis, polyneuritis, tabes, multiple myeloma, carcinoma, sarcoma, tuberculosis, osteitis deformans, senile osteoporosis, sometimes arthritis deformans, must be excluded.

It is not necessary to speak in detail of each possibility. Only complete and usually repeated examination can prevent error. Multiple myeloma can probably not be positively differentiated in all cases. Coxa vara, genu valgum, or rupture of pelvic articulations may have to be considered. Hysteria sometimes causes a striking simulation of osteomalacia. The differentiation of late rickets and osteomalacia depends upon the presence or absence of the rachitic epiphyseal changes, the less severe pain, and the tendency to cranial changes. Skiagraphic examination will often be of value. In some cases the results of treatment may aid.

**Treatment.—Prophylaxis.**—Gross errors in hygiene are important to correct in all endemic foci of disease, or in an individual case. This is especially important in the latter in periods of remission or after recovery. Fresh air, good food, exercise, and bathing are all useful. Nursing must not be permitted to an osteomalacic mother.

**Diet.**—Substances containing considerable lime and phosphorus should be preferred, such as milk, eggs, fresh meat, fish, beans, peas, and cereals.

**Drugs.**—Lime has been recommended, but is useless theoretically and has failed in practice. Calcium phosphate is but little better. The most useful drug is phosphorus, which is recommended by all who have used it persistently in large doses. It is given most frequently in cod-liver oil, in solutions of 0.06, 0.08 or 0.1 to 100. A teaspoonful of this is to be taken once daily, or 3 to 5 mg. (grain  $\frac{1}{20}$  to  $\frac{1}{12}$ ) at each dose. If cod-liver oil is not well borne, almond oil can be used, as in the *Oleum phosphoratum* of the British Pharmacopœia. It can also be taken in pills (*Pil. Phosphori*, U. S. P.) containing 0.6 milligram (grain  $\frac{1}{1000}$ ) each. The treatment must be continued at least one or two months. Improvement may not be apparent for three or four weeks. Kozminski recommends prolonged intermittent treatment based on the bone symptoms. Sternberg has given as much as 2.25 grams of phosphorus without toxic symptoms.

Phosphorus is slower and less certain, but safer than castration, the application of which we owe to Porro, who in 1878 reported a case of osteomalacia that recovered following the operation named after him.

Fochier and Levy thought sterilization was the cause of the recovery. Fehling (1886) showed the value of removal of the ovaries, and developed the theory already mentioned. While the theory is not accepted, the beneficial results of operation cannot be questioned, and although efforts have been made to attribute the recovery either to the narcosis or the operation *per se*, they have not been successful. The results of castration are sometimes remarkable. Pain sometimes disappears in a few days. But the improvement is neither uniform nor permanent, and phosphorus treatment may be carried out after it with advantage.

The indications have been formulated by von Winckel and others as follows: (1) Castration is indicated when other methods of treatment have failed and softening of the bones has advanced so far that life is endangered, or if the condition is such that death may occur before medicinal treatment can have an effect. (2) It is indicated in patients with pelves so narrow that pregnancy would necessitate Cæsarean section, and in case Cæsarean section is necessary.

Senator has used oöphorin with good results, but others (Latzko, Schnitzler, Bernstein) who have tried the method have failed. Bossi has advocated the use of adrenal preparations subcutaneously. There is an error regarding dosage in Bossi's publications, but he apparently used the material to the point of tolerance, as shown by chills, tremor, a feeling of suffocation, and a sense of constriction behind the sternum, coming on soon after the injections and lasting about a half hour. The symptoms increase with the number of injections. Besides Bossi, Ruffi, Tanturri, and Reinhardt had favorable cases, but others have failed.<sup>1</sup> Hoffmann has used the serum of thyroidectomized sheep and reports improvement in two months. Extracts of the pituitary body have been used with favorable results.

In a disease like osteomalacia any treatment based upon probable causes seems legitimate, but the chronic and variable course of the disease suggests the value of caution in announcing results.

**Symptomatic Treatment.**—In all cases various symptoms are certain to demand treatment. Comfort, the relief of pain, and the diminished danger of fractures should be provided for by a proper bed and care in moving or being moved. The skin requires care at all times, and especially if bed-sores threaten or occur. The bed pan and urinal must be used. Pain in the muscles and cramps should be relieved by hot baths, massage, fomentations, or at last morphine. Wine of colchicum seeds has been used by many with apparent advantage for the rheumatic pain. Diarrhœa and constipation must be treated as in other diseases.

<sup>1</sup> See the review of R. de Bovis, *La Semaine Médicale*, 1908, 241.



## CHAPTER XXVII.

ACHONDROPLASIA. HYPERTROPHIC PULMONARY OSTEO-ARTHROPATHY. PAGET'S DISEASE. OSTEOGENESIS IMPERFECTA. OSTEOPSATHYROSIS. LEONTIASIS OSSEA. MICROCEPHALUS. FACIAL HEMIATROPHY.

By CHARLES P. EMERSON, M.D.

### ACHONDROPLASIA.

**Definition.**—Achondroplasia is a disease of fetal life, possibly also of early infancy and childhood, which affects especially the long bones and the base of the skull. Its most conspicuous clinical features are: nanism, due to shortness of the extremities, especially of their proximal segments; large head; unusually deep depression of the root of the nose and trident-shaped hands.

**Terminology.**—Among the titles under which this condition has been described are: fetal rickets, intra-uterine rickets, congenital rickets, annular rickets (used also for other conditions), and micromelic rickets.

Achondroplasia<sup>1</sup> is, if we regard priority, the correct term, for under this title the condition was first described as a clinical entity. It is, however, an unfortunate name since cartilaginous bone formation is not, as the term implies, absent, but retarded. Nor is all cartilaginous bone formation retarded; for instance, the development of the vertebræ and short bones is little if any affected. For these reasons many prefer the term chondrodystrophia fetalis (Kaufmann<sup>2</sup>).

**Pathology.**—**Achondroplasia of the Stillborn.**—The great majority of achondroplastic fetuses die *in utero*, and of those which come to term the majority die soon after birth. These infants at once attract attention; the trunk and head are large but the limbs are ridiculously short. The umbilicus may be at the juncture of the middle and lower thirds of the body length. The chest is broad, the abdomen prominent. The nose is broad and flat; its root is depressed. The fat cheeks almost close the eyes. The tongue often projects from the mouth. But what attracts attention above all else are the short, often fin-like limbs, which give the child the appearance of a "marine mammal," a "cetacean," more than of a man. While the shortness of the limbs is seen in both distal and proximal segments, yet it is the proximal segments which are especially affected. These infants are plump, since there is present usually an abnormal amount of subcutaneous fat. The integument hangs in folds,

<sup>1</sup> Parrot, *Soc. d'Anthrop.*, 1878, p. 280.

<sup>2</sup> The best study of the pathology is that of Kaufmann, *Untersuch. über die sogenann. fetal Rachitis (chondrodystrophia fetalis)*, 1892.

giving the child the appearance of a "dwarf in much too large clothes." The genitalia are well developed.

The chief lesions of achondroplasia are in the long bones and seem secondary to a disease of their epiphyseal cartilages which inhibits their growth in length. The subperiosteal bone formation, on the other hand, seems normally or even abnormally active. As a result the long bones are thick and strong, but short. Achondroplasia may begin at any time before birth. The earlier it begins the shorter are the bones, even but one-half of normal length. When it appears late the bones may be of normal length. The bones most affected are the humerus and femur, next the tibia and ulna, and then the base of the skull. Because the fibula and radius seem little affected the disease is assumed to begin as a rule at or before the eleventh week of embryonic life. The diaphyses of the long bones are about normal in diameter but their walls are often thicker and harder than normal, and are sometimes even eburnated. They may have no medullary canal, or, as a result of bone absorption an abnormally large one. The smaller and more crooked the bones, the harder they are. The bones are very curved but symmetrically so. This is due to exaggeration of the normal curves which sometimes describe even right angles. In Lecadre's case<sup>1</sup> the legs were so bowed that the soles of the feet exactly faced each other. There are no bizarre deformities. The prominences for muscle attachments are hypertrophied. The epiphyses are usually of almost normal size, but sometimes are much swollen and misshapen. Some are shaped like mushrooms and give the bones an hour-glass appearance. In many cases the epiphyses are soft. They always are unusually vascular. Ossification in some cases is very retarded, in others is abnormally advanced, in which cases the epiphyses are firmly united to the diaphyses.

Those bones which do not pass through a cartilaginous stage in their development, for instance the frontal and parietal bones of the skull, grow to normal dimensions. This is true also of the clavicles, the pelvis and the ribs. The short bones—the vertebræ, metacarpals, etc.—develop from cartilage and are said to escape the disease; but it is quite certain that they do not always (if ever) entirely escape, although their involvement may be slight.

The skull is voluminous and suggests hydrocephalus. The parietal bosses and the forehead are prominent. In some infants true hydrocephalus has been present; in others the unusual thickness of the bones explained in part the large size of the head. In still other cases the skull is abnormally fragile. Two such cases were reported—one a seven and a half months' fetus reported by Champetier de Ribes and Constantin-Daniel—whose heads burst during delivery "without violence" by the Mauriceau method. In the case reported by Mary A. Smith no bone had developed in the vertex of the skull. However, the diagnosis in both these cases is doubtful. (Was lues present?)

The frontal and parietal bones usually develop normally, but not so the base of the skull. At birth the foramen magnum is surrounded by a ring

<sup>1</sup> *Paris Thesis*, 1856.

of four separate bones, which normally are not united to form the occipital bone until the sixth year. Provision is thus made for an increase in the diameter of this foramen. In the achondroplastic stillborn infant a premature synostosis of these bones is usually found, and so the funnel-shaped foramen magnum is surrounded by a ring of solid bone, which is incapable of much increase in diameter. The pressure which this small and solid ring must exert on the growing cord is thought to explain the premature death of these infants.

The portion of the base of the skull formed by the occipital and sphenoid bones, the "*os tribasilaris*," consists at birth of three pieces, the basilar portion of the occipital bone and the presphenoid and postsphenoid bones. The synchondrosis intersphenoidalis between the last two bones closes at about the time of birth, but the cartilage uniting the sphenoid and occipital bones is not solid until about the eighteenth or even the twenty-fifth year. In the achondroplastic stillborn infants these bones are often firmly united, and the clivus deprived of the possibility of growth in length. The clivus is in these cases abnormally vertical (*Virchow's kyphosis of the skull*). This shortening of the base of the skull from the normal 7.6 cm. to 4.6 cm., or even less, explains, according to Virchow, the depressed root of the nose. Kassowitz found that the root of the nose could be depressed also when the tribasal bone was of normal length or even wholly cartilaginous, and he thinks that the tissues anterior to the sphenoid bone in part at least explain the profile of the face. But the fact that this bone is cartilaginous does not indicate that it is capable of further growth.

The ribs show a well-marked rosary, but the prominences are not due, as in rickets, to hypertrophy of the cartilage, but to overgrowth of the bone in a cup over the end of the cartilage. In some cases (Grawitz) the rosary is only apparent, and due to an angular deformity at the costochondral articulation. The rosary is usually much more prominent on the inner than on the outer aspects of the thorax.

The vertebræ sometimes show a premature synostosis of the body and arch of the upper cervical vertebræ which thus develop in a way similar to that modified vertebra which we call the occipital bone. The clavicles are usually normal. Shattuck was the first to describe premature synostosis of the coracoid process of the scapula. The pelvis is sometimes distorted, massive, and ill-shaped, and the sacrum is tilted forward.

Microscopic examination of the epiphyses reveals at once the explanation of the deformities. The epiphyseal cartilage is abnormal, both its ground substance and its cells. The line of ossification, often not visible without magnification, is quite straight but very narrow. The zone of proliferation of the cartilage cells containing the columns of proliferating cells is absent. The cells are irregularly distributed, few and scattered. This aplasia of the zone of cartilaginous bone formation is held to be the characteristic lesion of achondroplasia. The intercellular substance of the epiphyseal cartilages is decidedly abnormal. Near the surface of the epiphyses it is transformed to fibrous tissue; the rest, instead of being hyaline, is often mucoid and sometimes fibrillar. This fibrillar structure may be so marked that its network can be easily seen. The cartilage is

sometimes riddled with vacuoles. There is sometimes a broad mucoid or gelatinous zone between cartilage and diaphysis. These changes are so marked that Trippier said: "There are no epiphyses. In their place is another tissue, reddish and completely resembling muscle tissue." In other cases the epiphyseal cartilage comes in immediate contact with a well-ossified spongiosa, or is separated from it by a thin zone of swollen cells. In some cases the completely calcified laminae of spongiosa show all the signs of metaplastic ossification of the cartilages. So abnormal are the epiphyseal cartilages that the wonder is that the bones grow as long as they do (Kassowitz).

But the most interesting lesion of the epiphyseal cartilages is the growth from the periosteum of fibrous tissue which invades the cartilage. The amount of this invading tissue varies from a few fibres to a definite membrane which seems to push itself in between diaphysis and epiphysis and so by checking enchondrial bone formation inhibits the growth in length of the bone. This band of fibrous tissue is vascular and encloses in its meshes islands of cartilage and also fat cells, evidence of its origin from the periosteum. It is always thinnest in the centre and thicker at the margins. If this tissue invades the cartilage from but one side, the growth of bone is checked on this side only. Thus arise many of the angular deformities which the bones of achondroplastic skeletons show and, Kaufmann thinks, many of the so-called fractures. One of the best descriptions of these changes is that of Urtel.<sup>1</sup>

In nearly every case the epiphyseal cartilages are very vascular. The vessels arise in the perichondrium and invade the cartilage, running at right angles to the shaft of the bone. Along their course especially does the fibrous tissue develop. The marrow spaces are sometimes large and the marrow tissue is very cellular and vascular. Giant cells are numerous and by their arrangement indicate active bone absorption, which probably explains the osteoporosis interna which one often sees. This bone absorption may outstrip the active periosteal bone formation and so the hard cortex be reduced to so thin a shell that it fractures easily.

Lesions of other organs than the bones are inconstant. In a few cases a mild degree of true internal hydrocephalus was described, but Durante<sup>2</sup> thinks there is no well-marked nervous lesion. The thyroid gland of the majority of reported cases was described as normal. In a few cases the thyroid was large, "twice the normal size," "hypertrophied" (Salveti), or "absent." The thymus was large in a few cases. The liver has several times been reported as enlarged.

**Varieties.**—Kaufmann attempted to classify the thirteen infants he studied in three groups; the hypoplastic, the malactic, and the hyperplastic. In the first group, "*chondrodystrophia fetalis hypoplastica*," the five cases had abnormally short but stout trunks, large heads, and very short extremities, over which the investment of soft tissues hung in tumor-like folds. The thorax was narrow above, wide below, and the "rosary" was very marked. The root of the nose was much depressed.

<sup>1</sup> Halle Thesis, 1873.

<sup>2</sup> Bull. de la Soc. Anat., 1900, p. 785.

The eyelids and the lips were swollen, and the cheeks thick. The epiphyseal cartilages were normal in size, firm in consistency and very vascular. In the second group, *chondrodystrophia fetalis malacica*, were three cases. The noses of these infants were depressed as a whole, and the epiphyseal cartilages were abnormally soft. Of the third variety, *chondrodystrophia fetalis hyperplastica*, Kaufmann reported but one example, which he believed to be the only case of the variety up to this time reported. This case differed from the others in that the mushroom-shaped epiphyses were soft and very vascular and caused a swelling of joints, as in rickets. The growth of the cartilage is described as "wild." The bones were hard and seemed disposed to fracture; in fact five long bones showed fresh breaks. The trunk of this infant was unusually long (42 cm.). Kaufmann admitted that four of the thirteen cases he described could not be classified in any of these three groups. Regnault<sup>1</sup> denied that the cases can be thus classified. He considered all as grades of one and the same disease, those children only with the hypoplastic variety being viable.

Cases of partial achondroplasia have been reported. Pauly and Teyssier<sup>2</sup> reported the case of a man, 138 cm. in height, who lacked the head abnormalities, the trident hand, and the relatively long fibulae. Regnault also is confident that partial forms do occur.

Achondroplasia is often combined with other deformities and abnormalities. Among these are hypospadias, cervical cysts, spina bifida, arrested development of the ears, arrested development of the ensiform cartilage, umbilical or inguinal hernia, cleft palate, genu valgum, and a large variety of tumors.

**Achondroplasia of Children and Adults.**—Our knowledge of the pathology of achondroplasia of children and adults is obtained chiefly from the study of skeletons and of radiographs of living cases. A few cases have come to autopsy. The skull is voluminous, its parietal and frontal bones are usually prominent and its bones sometimes very thick. The foramen magnum is small. Its dimensions in the three adult skeletons studied by Breus and Kolisko were: 1.9 by 2.5 cm., 2.2 by 2.8 cm., and 2.9 by 3.4 cm. These writers suggest that cases of achondroplasia be divided into two groups, those with the premature synostosis of the bones surrounding this foramen and those without. In some cases the skull seems unaffected. The ribs show a bony rosary. The vertebrae seem affected in severe cases, but not in the milder ones. The arches and bodies of the upper cervical vertebrae in some seems to have united prematurely, if we may judge by the stenosis of the spinal canal of certain skeletons. The long bones of the extremities are short and thick, their normal curves are exaggerated and the protuberances for muscular attachment are prominent and strong.

Studies of the *radiographs* of adults with achondroplasia suggest two distinct groups of cases—those in which the epiphyseal line persists even at forty-five years of age, and those in which it disappears abnormally early. In cases of the first group the ossification of the epiphysis is much retarded (the hypoplastic form of Variot, the true achondroplasia of

<sup>1</sup> *Arch. gén. de méd.*, February, 1902.

<sup>2</sup> *Prov. méd.*, xiv, p. 489.

Parrot). In the second the ossification is abnormally advanced, and therefore the epiphyseal cartilages are much reduced in thickness (the hyperplastic form of Variot). Variot points out that in both cases the result is the same; that is, the long bones do not increase much in length. As an illustration of the hypoplastic variety, he reports the case of a girl, aged thirteen years, in whom ossification was much retarded. The head of the humerus seemed crushed, and the lower end of this bone was cartilaginous for 1 or 2 cm. above the joint. The lower ends of the radius and ulna were little ossified, while the carpals and especially the metacarpals were almost as cartilaginous as at birth. The same was true, but to a less degree, of the bones of the lower extremities. Variot contrasts this case with that of Méry and R. Labbé, which he considers, illustrates the hyperplastic form. Their patient was a boy aged twelve years, in whom ossification was abnormally advanced and the epiphyseal lines early obliterated. Since the head of the girl he reported was unaffected, while all the other signs of achondroplasia were present, Variot proposes to call such cases "achondroplasia without cranial involvement," or "epiphyseal dyscrasia."

The humerus of the adult cases is surprisingly short. In ten skeletons, all apparently of adults, the humerus measured in length from 14 to 17 cm., average 15.3 cm. (normal length about 27 cm.); the ulna, from 10.7 to 15.8 cm., average 13.2 cm.; the radius, from 8 to 14 cm., average 11.4 cm.; the femur, from 16.5 to 24 cm., average 20.7 cm.; the tibia, from 11.6 to 20 cm., average 16.3 cm.; and the fibula, from 16.2 to 21.2 cm., average 19 cm. The skeleton of the hand measured from 10.3 to 14.5 cm. (normal 20 cm.), and of the foot, from 14.5 to 18 cm. (normal 25 cm.). The scapula is smaller than normal, especially in its vertical diameter, and its glenoid cavity is apparently too small for the head of the humerus (Cestan). The olecranon seems to be too large for the olecranon fossa. The fibula presents an interesting study for it is always much less affected than is its companion tibia, but since it is "splinted" to the tibia it must find room for its greater length by abnormal curves or by extending as high or even higher than the tibia and entering into the formation of the knee-joint.

**Achondroplasia in Art and History.**—Of all dwarfs only those with achondroplasia and rickets could serve as the court buffoons so popular at the end of the seventeenth and again in the eighteenth centuries. That achondroplasics would have been particularly well adapted for this purpose is seen now by the number now found on the vaudeville stage. They are quick-witted, very active, and enjoy this occupation.

The diagnosis of achondroplasia in products of art is not easy, for the artist seldom portrays the fine points necessary for a positive diagnosis. In a doubtful case it would be difficult to distinguish in a picture between a myxœdematous and an achondroplastic dwarf, for both are only exaggerations of the fetal type, which the artists and sculptors of antiquity evidently used for symbolic purposes. The normal infant has a relatively large trunk and head and short extremities. In various museums, we were surprised at the number of crude wood and stone images, from a variety of barbaric tribes, which were micromelic. Pictures

on the coffins of certain mummies suggest that these mummies were achondroplastic dwarfs. In more recent art, Velasquez's "Sebastian de Morra" and "El Primo" and some of the dwarfs in the pictures of P. de Veronese and Julio Romano are almost certainly achondroplastic.

**Etiology.—Heredity.**—This is interesting not only for its own sake, but also because of the bearing it may have on the origin of certain races of dwarfs and of certain varieties of animals (granting of course that animals do have this disease). Among the races of dwarfs are the Akkas and other African tribes, the Mincopies of the Andaman Islands, the Semangs of Malacca, and the *Ætas* of the Philippines. It is worthy of mention that there are no white races of dwarfs, and that all white dwarfs are evidently pathological.

That achondroplasia can be inherited there is now no doubt. Marie accepted three cases as certainly inherited (Porak's, Baldwin's, and Boeckh's). One of the most interesting cases on record is reported by Porter.<sup>1</sup> The father, aged eighty years, and his two sons were achondroplasics. Also the father and a brother of this octogenarian were said to be achondroplasics. This would mean six cases in three generations. The mothers of these patients were all normal women. Comby<sup>2</sup> reports a boy and his maternal grandfather who were achondroplasics, and Poncet a brother, sister, father, and grandfather.

Achondroplasia is also a family disease appearing more than once in the same generation. Good illustrations are the family reported by Porter and the brother and sister reported by Lannois. Chavigny's patient said he was the tallest of four children, all shaped like himself. Chiari<sup>3</sup> states that a man of normal size (but whose father was a dwarf) was the father by two normal sisters of two stillborn achondroplastic infants. Hutchison reports that of twins he saw the one was an achondroplastic girl and the other a normal boy.

It is claimed that achondroplasia occurs in animals also. As best illustrations are cited the rare "bull-dog calves," which have very short snouts, short legs, thick, wrinkled skin, and normal development of the short bones. Some claim that the dachshund, the pug dog, guinea-pigs, Ancon sheep, the Nato's cow of Chili, Yorkshire pigs, etc., are cases of achondroplastic animals, but the better opinion seems to be that the resemblance here is accidental.

**Gigantism.**—The question of a possible relationship between achondroplasia and gigantism was raised by Lannois who reported the case of an achondroplastic man, aged twenty-five years, whose next older brother measured 192 cm. in height. The interval between the births of these two persons was very short. Two other brothers were very tall. The father measured only 160 cm. in height. Lunn<sup>4</sup> reported a case of achondroplasia in a patient whose father measured six feet two inches in height, his mother five feet ten inches, and whose three brothers and one sister were normal. Claudius (Marie's case) came of a tall family. Gigantism is in many cases due to pituitary disease. This gland has been carefully

<sup>1</sup> *Brit. Med. Jour.*, 1907, i, 12.

<sup>2</sup> *Bull. de la Soc. des Hôp. de Paris*, 1902, 551.

<sup>3</sup> *Deut. med. Wchschr.*, 1913, xxxix, 438.

<sup>4</sup> *Trans. Clin. Soc.*, London, xl, 252.

studied recently in cases of achondroplasia coming to autopsy, but in only one case was it found enlarged or otherwise abnormal.

The numerous cases of achondroplasia which have occurred in large families suggests a possible relationship between numerous pregnancies, especially when these occur at short intervals, and this condition. Herrmann's case was one of nine children, Wood and Hewlett's was the thirteenth of sixteen children. Méry and Labbé's patient was one of thirteen children.

Congenital dislocation of the hip is sometimes associated with dwarfism and in not a few cases it has been associated with achondroplasia. Such cases have been reported by Kassowitz, Kirchberg, Simmonds, and Kaufmann. Grawitz suggests that congenital dislocation of the hip is due to achondroplasia limited to the pelvic bones, the lack of proliferation of the cartilage explaining the shallowness of the cotyloid cavities.

**Symptoms.**—While the great majority of achondroplastic infants are stillborn or die during the first year, those who reach their second year seem to have the average expectancy of life. As the child develops the disproportion between the amount of loose skin and the length of limb disappears. Teething, talking, and walking, the criteria by which an infant's progress is usually measured, seem normal in the majority of these children, but some, to judge from the records, are backward.

The adult achondroplastic presents a striking appearance, the most noticeable points of which are the short extremities, the body of almost normal size and the large head. Achondroplasias are dwarfs in the sense that they are shorter than normal men. Of eighteen achondroplastic men all over twenty-one years of age, the heights varied from 93 to 138 cm., averaging 119 cm. Of eighteen achondroplastic women all over twenty-one years of age, the heights varied from 97 to 130 cm., averaging 116 cm. The trunk of the achondroplastic adult is usually described as normal, but in well-marked cases there does seem to be a slight microsomia. The women are usually stout, while the men are very muscular. Micromelia is the important mark of achondroplasia; that is, the legs are short, even but half the normal length, and the arms are affected in about the same degree. This shortening is not symmetrical for the root segments, the upper arm and the thigh are relatively more shortened than are the distal segments; that is, the condition is one of rhizomelia. The upper extremities hang down about to the crest of the ilium, seldom below the greater trochanter. The shoulders are attached slightly more posteriorly than normal. Owing to the small size of the cotyloid cavity the arms cannot hang vertically, but project slightly from the sides (Marie, Poncet). The bones of the arms are only slightly curved. Since the olecranon cavity is small, the humerus and the ulna are articulated at an angle and the arm can seldom be extended straighter than 135 degrees. The articular fold of the elbow is more oblique than normal. The radius seems relatively too long and its head is so large that supination is limited.

The hands have a characteristic shape. They are small, short, and thick and, since the fingers are all of about equal length, have a cubical shape. The ring finger seems to lie in a plane posterior to the others



and is almost covered by them. The first phalanges lie parallel, but beyond these some or all of the fingers, as a rule, diverge from one another like the prongs of a trident, giving the hand the "trident shape" described by Marie. The legs are often bowed or knock-kneed, which is due not to curves of bones, but to the joints, for the bones are articulated at an angle. The head of the tibia is large. The marked curves and high position of the fibula are often quite evident on palpation. The toes are of almost equal length. The joints are abnormally lax, especially those of the knees, the hands, and the feet. The spine shows an interesting and quite uniform abnormality in that the sacrum tilts forward into the pelvis, with a resultant sharp lumbar lordosis, which explains the contraction of the pelvis. The thoracic spine is so straight that the back is flat.

*Macrocephaly* is the rule. The head is usually globular in shape, with well-marked frontal and parietal bosses. Apert reports a patient with a head 66 cm. in circumference and Marie a man whose head measured 67 cm. in circumference. Of eleven men, all over twenty-one years of age, the heads measured in circumference from 54 to 67 cm., averaging 59 cm.; in the case of five women this circumference varied from 52 to 56 cm., averaging 54 cm. Brachycephalia is an almost constant feature and the cephalic index is usually over 0.9; in one of Marie's patients it was 1. The shape of the head suggests hydrocephalus, but it is as a rule the thickness of the skull which explains this shape (Cestan). In most cases the closure of the fontanelles and sutures was retarded.

The face seems small, but actually is large. The features are coarse, the forehead prominent, and the eyes deep. The nose is quite characteristic. It is short, its base is broad, its root very deep, its tip prominent and thick and its nostrils large. The arch of the hard palate is frequently high, but the teeth usually show no abnormality. Not all cases show these abnormalities of the skull, and Variot proposed to separate them into a separate group, "achondroplasia without cranial dystrophy." The shape of the head may be asymmetrical (Langenbach).

The adult achondroplasias are remarkably strong, even when compared with normal men, and seem to choose, or at least not to avoid, strenuous work, even when elderly (Porter). They are excellent athletes, and popular circus acrobats, attracting attention not only because of their conspicuous build, but because of their strength and skill.

It is the general belief that these patients are rather limited mentally, are mischievous, intemperate, and lascivious. This is by no means the rule. We have seen one and several have been reported whose mentality was of the highest order, who were students and teachers of excellent ability. The child of the lower classes must depend on the public school for education, and here the achondroplastic child will learn little that is good, since his schoolmates will make his life miserable. It is interesting that the majority of the bright patients are women. Of some it is said that they were bright when children but mentally rather inferior as adults. This would agree with the idea that in some cases there is retarded intellectual development at puberty. Marie called attention to the probable relation between the height of the patient and his intellectual develop-

ment, a point well illustrated by three adult men in his clinic. Cladius, 107.5 cm. tall, aged twenty, was a child in behavior and mind and physically was evidently not at puberty. Anatole, aged forty-one, 122 cm. tall, was frolicsome, boastful, mischievous, intemperate, and lascivious. Surgeus, aged sixty-six, just a little taller than Anatole, was a jeweller by trade and a sober, married man. The age at which the disease develops possibly may influence the mental development. The children born achondroplastic appear to develop mentally as rapidly as do other children, but the mental development of those in whom the disease continues, or perhaps begins, after birth would seem to be slow.

Sexually, these patients are well developed both anatomically and functionally. Many patients of both sexes seem unusually salacious. Some men are notoriously so, while unmarried women in the obstetric clinics have given the best opportunities for the study of achondroplasia. Apert mentions a woman with contracted pelvis who underwent operations for three pregnancies.

One of the most interesting subjects connected with this disease is its postnatal course. And yet one should remember that achondroplasia itself is not a disease but a condition, a deformity, the result of a disease whose nature is still unknown and the date of whose course is uncertain. The first cases described were all stillborn children in whom the disease seems to have run its course. In most cases the disease may run its entire course and heal *in utero*; that is, at birth there is no evidence of active epiphyseal new-bone formation. These cases are stillborn. In other cases the disease seems still florid at birth, while it would seem as if some infants apparently normal at birth develop achondroplasia later.

Kassowitz<sup>1</sup> followed the postnatal course of the disease in several patients for years after birth, reporting measurements and publishing photographs which are very convincing. Both the mother and grandmother of Swoboda's<sup>2</sup> patient said that the limbs of this baby girl were of normal length at birth. She was stout and so soft and flabby that they feared to lift her. She had a congenital left genu valgum with flat foot. Before she was ten years old, at which age the diagnosis of achondroplasia was made, at least forty medical examinations had been made by almost as many physicians. Her head began to enlarge after the first year, and she walked at three years. At first there was lumbar kyphosis, which changed to a lordosis as soon as she began to walk. The muscular system was well developed. The soft, tender flesh and the excessive sweating about the head led to the diagnosis of rickets, which was the opinion until she was ten years old. Intellectually this girl was normal.

In many cases these children suffered during infancy from some uncertain acute illness, following which in some the achondroplasia was first noticed. "Couldn't hold up his head," "hydrocephalus," "lost for a while the use of the limbs," "an acute illness," "was weak," are among the expressions used to describe these illnesses. The writer had excellent opportunities to study one marked case, an adult man, whose parents were by no means poor and ignorant, whose mother has

<sup>1</sup> *Wien. med. Woch.*, 1902, No. 28, 1358.

<sup>2</sup> *Wien. klin. Wchschr.*, 1903, No. 23, 669.

repeatedly told the writer the story of his infancy. There seems no doubt that the child was born with normal proportions, had, as an infant, an acute disease resembling rickets, and that signs of his deformity began at about four years of age.

We may, therefore, believe that achondroplasia is a condition which may develop after birth, or rather that the deformities of achondroplasia appear after birth, for it may perhaps be unwise to confuse these deformities with the disturbance of which they are the result. The symptoms of the acute disease remarkably resemble rickets. There is retardation of development, flabbiness and tenderness of the flesh and sweating of the head. The teething, talking, sitting up, and walking are all retarded, and the relative micromelia is first noticed after this illness.

**Nature.**—All achondroplastic infants were formerly referred to as “monstrosities,” and the adults as “dwarfs.” Virchow termed the infant he reported a “fetal cretin,” thus attributing the deformity to lack of development of the thyroid gland. Fortunately this infant was preserved as a museum specimen and so was studied later by Klebs.<sup>1</sup> There is no doubt but that this infant was typically achondroplastic. In several other achondroplastic infants lesions of the thyroid were recorded. Many cases have been tested with large doses of thyroid extract, but without improvement. Cavazzani reported a case which he thinks unique. The mother of an achondroplastic infant had exophthalmic goitre. During pregnancy she took large and finally toxic doses of thyroid extract. Later the child was given the same drug in large doses. Both showed a marked tolerance to this extract.

Against the thyroid theory it is claimed that in achondroplasia ossification is rapid and in cretins retarded. But this is not always true. Simonds stated that there certainly are cases with combined achondroplasia and myxœdema. In their recent paper Symmers and Wallace<sup>2</sup> uphold this view. They maintain that there is a cretinoid variety of chondrodystrophia fetalis in which in addition to changes in the osseous system there are modifications in the soft tissues (lips, cheeks, eyelids, nostrils, ears, tongue, etc.) which are attributable to pathological defects in the thyroid gland. In these cases they found extensive chronic productive inflammatory process of the thyroid gland eventuating in replacement of the colloid vesicles by means of overgrowth of alveolar epithelium or by invasion or substitution of the alveoli by connective tissue elements derived from the interstitium, aided in both instances by compression exerted from the outside by the contracting fibrous trabeculae. It may be, however, that such changes in the thyroid gland are not the cause of the achondroplasia, and if they are we must then explain the thyroid lesions. It is possible that we may find them the result of the same disease which disturbed the cartilaginous bone formation. This same point of view should be held in discussing the lesions found in other ductless glands, for Cestan claimed that the disease certainly originates *in utero* before glandular activity begins, and so cannot primarily be due to any defective internal secretion.

<sup>1</sup> *Arch. Exp. Path.*, 1874, Bd. ii, p. 70.

<sup>2</sup> *Arch. Int. Med.*, 1913, xii, 37.

H. Müller (1860) indentified this condition with rickets. The ordinary rickets, he said, develops after birth; this form runs its whole course *in utero*. Since both rickets and achondroplasia are conditions as yet not understood, we are in no position to say whether or not they are related. They may indeed be manifestations of one and the same disease. They surely are no more different than various forms of tuberculosis, for instance. The three questions are: Do the two diseases resemble each other clinically and morphologically? and the almost universal answer is that they do not sufficiently to justify the term "fetal rickets." The second question: Is there a fetal form of the disease we now term rickets? The cases now termed fetal rickets would seem to be different from achondroplasia, and yet the more detailed the description the more certain we are that they are cases of achondroplasia. The third question: Is there a postnatal form of achondroplasia and does it resemble common rickets? There surely is and it does not. Fetal rickets is described as a disease beginning in the second half of pregnancy and in full bloom at birth. Its lesions are isolated and circumscribed, consisting of bony softening and tumefaction, and which produce asymmetrical curves. The ribs are always affected. The earlier writers separated fetal rickets, which runs its entire course *in utero*, congenital rickets, florid at birth, and common rickets, which begins after birth.

In both rickets and achondroplasia the cartilages are abnormally vascular, but the vessels differ in size, structure, and arrangement. In rickets there is luxurious cartilaginous formation. The columns of swollen cartilage cells are abundant and extend in all directions. In achondroplasia they are scanty, and (differing from rickets) the ground substance of the cartilage is nowhere normal but fibrillar or glassy. In postnatal rickets there is very little real shortening of the bones. The limbs are short because the bones are bent, and the bends are pathological, asymmetrical and bizarre, while in achondroplasia they are symmetrical, and only exaggerations of the physiological curves. In rickets there is relatively more involvement of the bones of the thorax, pelvis and spinal column than in achondroplasia, although it is now granted that in the latter disease these bones may be slightly affected. There is a rosary in both cases, but careful examination of the bones shows the marked difference that in rickets the swelling is cartilaginous, in achondroplasia it is bony. In rickets the shape of the head is quite different from that in achondroplasia, although we have seen an achondroplastic person with typically rickety head. Even since the first it has been granted that typical achondroplasias do show signs of rickets, *e. g.*, the high forehead with prominent frontal bosses, Harrison's groove, and some bizarre bends of the long bones. But we have contrasted above achondroplasia and common rickets, two conditions neither of them a disease, and both the results of unknown diseases. We have brought forward no evidence against the possibility that the two conditions are different forms of one and the same disease. Marie admits the occurrence of both in the same patients, and Macewen<sup>1</sup> emphasizes the occurrence of both diseases in different children of the same family.

<sup>1</sup> *Brit. Med. Jour.*, 1907, ii, 1646.

Symington and Thomson<sup>1</sup> described achondroplasia as an arrest, or perversion, of the normal processes of endochondrial ossification of the most definite and universal character, involving, during intra-uterine life, every element of the skeleton. Durante<sup>2</sup> described the process as a sclerosis of the zone of endochondrial ossification.

It is claimed that achondroplasia is the result of a maternal intoxication or a fetal auto-intoxication. Durante<sup>3</sup> reported one patient whose mother was luetic, and another whose mother died of liver and renal trouble. He reports also<sup>4</sup> a patient whose mother died of tuberculosis during the pregnancy. Peloquin believed it due to a maternal infection. The opinion is gaining ground that achondroplasia is a luetic or paraluetic affection. While the question of the relation to rickets is at present not capable of solution, that of lues is, and in all stillborn achondroplastic infants search should be made for the *Treponema pallidum*.

Syphilis can produce a micromelic dwarf. Birrenbach<sup>5</sup> reported the case of a stillborn luetic infant which he says was certainly a case of achondroplasia. The coincidence of the two conditions may have been accidental.

**Diagnosis.**—For the diagnosis of achondroplasia of the stillborn child Durante believes that the gross appearance is not enough but microscopic examination should be made to demonstrate disturbance in the cartilaginous ossification. In the living child the radiographs may show either too little or too much cartilage at the epiphyses.

For the diagnosis in the adult it will be necessary to state first just what features the case must present to warrant the diagnosis of achondroplasia. Most will agree that these are dwarfism due to rhizomelic micromelia in an individual whose trunk is of almost normal size. But is this clinical picture always due to the same disease or can several diseases cause it? There are several types of dwarfs. Some are small in all dimensions but have practically normal proportions. Lanceraux gives the upper limit of height of these "true dwarfs" as 120 cm. In this strict sense the achondroplastic individual is not a dwarf. Other dwarfs owe their condition to disease of the skeleton, especially rickets and achondroplasia, and still a third group to general disturbance of nutrition, as myxœdema and congenital lues.

*Infantilism* is a term frequently used of dwarfs. By this is meant a condition in which the physical proportions of youth persist in adult age. The growth in height may not be stunted, although it usually is, the limbs are relatively long, the head is small, and the secondary sexual characteristics are late in development. It is at once evident that the term is a misnomer. Puerile would be a better term. The infant is in this sense not "infantile," but fetal, for its limbs are relatively short and the head large. The achondroplastic dwarf resembles much more the infant than the youth, but the secondary sex characteristics are well developed, even abnormally so. There is a rule which will be found useful

<sup>1</sup> *Laboratory Reports of Royal College of Physicians*, 1892, iv, 238.

<sup>2</sup> *Rev. méd. de la Suisse romande*, 1902, 809.

<sup>3</sup> *Bull. de la Soc. Anat.*, 1900, p. 785.

<sup>4</sup> *Rev. méd. de la Suisse romande*, 1902, p. 809.

<sup>5</sup> *Thesis, Griefswald*, 1901.

in studying patients for traces of infantilism or achondroplasia. In the normal adult the length of the forearm from the tip of the olecranon to the tip of the middle finger is equal to that of the lower leg from the lower tip of the patella to the sole of the foot, and this distance is approximately twice the height of the head from the level of the vertex to the point of the chin. The achondroplastic patient should be compared always with a person of the same age, not with a child of the same height. This is important, since Cestan has shown that many illustrations in art may not be cases of achondroplasia, but of infantilism (fetalism).

Jeffrey Hudson grew over two feet after he was thirty years of age. Borwiloski was twenty-eight inches (71 cm.) when twenty-two years old, but grew even when in extreme old age. Bébé, the dwarf of King Stanislaus of Poland, died when twenty-two years of age and 90 cm. in height. Infantilism is seen in cases of mitral heart disease, but no cardiac dwarf has as yet been reported whose height was under 140 cm. Infantilism is seen also in idiots and in cases of pancreatic disease. But these cases seldom confuse, for their slender trunk, lack of development of the secondary sexual characteristics and especially their small head, will distinguish them from achondroplastic dwarfs. The achondroplastic person is a deformed dwarf, and this deformity consists in rhizomelic micromelia. That is, he is an "ectromelus" ("aborted limbs," a monster with arrested development of the limbs). He also suggests a "phocomelus" ("seal limbs," a monster with shortened arms and thighs, with hands and feet attached almost directly to the trunk). The adult rachitic dwarf is short chiefly because his legs are bowed. Some micromelia is present but not rhizomelia. His arms are relatively long, the thorax is much involved, and the hands are not trident-shaped. The myxœdematous dwarf or cretin is distinguished by the infantile condition of his body, trunk and limbs, his very reduced mental state, his dry and myxœdematous skin, dull facies, etc. Patel considers that dwarfs with practically normal proportions and without signs of myxœdema or achondroplasia are cases of acquired athyroidism due to atrophy of the thyroid gland at the age of ten or twelve years.

**Prognosis.**—The great majority of achondroplastic infants are stillborn, having died at the eighth month of intra-uterine life. The reason for the death of so many at just this age may be due to pressure on the spinal cord by the prematurely synostosed occipital bone. Regnault believed that of achondroplastic infants only those with the hypoplastic form survive, and by hypoplastic he meant sluggish cartilaginous bone formation.

**Treatment.**—Any treatment for achondroplasia will, of course, help only those patients in whom the disease is florid after birth. For these, careful feeding, massage, passive movements, electricity, and all possible measures to keep up the general nutrition are indicated.

**Congenital Osteosclerosis.**—Schmidt<sup>1</sup> reported the case of an infant who died thirty hours after birth, with slight thickening of the long bones but considerable thickening of the spongiosa, especially of the

<sup>1</sup> *Verh. d. Deut. path. Gesellsch.*, 1907, ii, 288.

vertebræ, base of the skull, sternum, and the ends of the long diaphyses. The hypophysis of this infant weighed 0.28 gram, or twice the normal weight for an infant of this age. There were no signs of lues. Schmidt was sure that the process was not due to marrow changes. He quotes Assmann<sup>1</sup> who reported such a case, and ascribed it to healed intra-uterine leukemia. The hypophysis in this case was small but very vascular.

**Pseudo-achondroplasia, Periosteal Dysplastic Type.**—Gonnet<sup>2</sup> reported a case of "pseudo-achondroplasia of the periosteal dysplastic type." In body form this child resembled an achondroplastic infant, but crepitation of the humeri, femora, and tibiæ could be produced, and the membranous pouch of the skull contained only a few islands of bone. Trillat<sup>3</sup> reported a similar case with quite typical achondroplastic body form, but with many fractures of the long bones and ossification of the cranium.

**Dyschondroplasia.**—This is the name Ollier<sup>4</sup> applied to cases with irregularity in the ossification of the bones. The seemingly normal cartilage ossifies abnormally slowly. Radiographs of these cases show transparent areas in the bones, which at first glance might suggest sarcoma. This condition is seen in children. They are not dwarfs, and if they are micromelic it is not to such a degree as in achondroplasia. They have not the skull changes and seldom the trident hand of achondroplasia. There may be slight atrophy of the affected limb.

**Hyperchondroplasia.**—This is a condition with abnormal lengthening and attenuation of the bones, with increase in width of cartilages of conjugation. Méry and Babouneix<sup>5</sup> reported such a case. Fournier had reported such cases as due to hereditary lues.

## THE HYPERTROPHIC OSTEOPATHIES.

From the large and very confused group of the hypertrophic osteopathies certain types have been separated as distinct diseases. Among them are: general hyperostosis (Friedreich, 1868); osteitis deformans (Paget, 1876); acromegaly (Marie, 1885); chronic pulmonary osteoarthropathy (Marie, 1890). And yet of all the many cases of hypertrophic osteopathy but few can definitely be assigned to any one of these subgroups. Some are reported as atypical cases, others as "mixed" cases, others as separate diseases, while more were atypical probably only before the disease was fully developed and lost their peculiar features years after they were reported.

While most writers consider the above diseases as distinct, several French writers insist that they all are illustrations of one or perhaps two different diseases, and that their chief differences lie in the localization of the lesions. Some think that the causal disease is tuberculosis; others, syphilis. Pathologically there is a certain similarity in all these cases; one finds bone absorption and new bone formation; callus building

<sup>1</sup> *Beit. z. osteoschl. Anämie, Zeigler's Beiträge*, 1907, Band xli.

<sup>2</sup> *Bull. de la Soc. d'Obstet. de Paris*, 1908, xi, 185.

<sup>3</sup> *Ibid.*, 183.

<sup>4</sup> *Bull. et mém. de la Soc. de Chir. de Lyon*, 1889-1890, iii, 22; Molin, *Thesis*, Lyon, 1901.

<sup>5</sup> *Soc. Hôp. de Paris*, July 4, 1902.

and condensing osteitis; spontaneous fractures and various deformities, either angular, resulting from one area of local softening or fracture, or general, from a bending of the bone as a whole. Among the earlier terms used for these cases are rickets, osteomalacia, osteoporosis, osteosclerosis, etc.

**Paget's Disease; Osteitis Deformans; Ostitis Deformans.**—The term *ostitis deformans* has long been used of various bone diseases which cause deformity of one or several bones. This term included not only nearly all the constitutional diseases of bone as we now know them but also callus building in the repair of genuine fracture, of spontaneous fractures in lues, congenital deformities, etc. The cases now described as of Paget's disease were formerly certainly grouped under this title (Sternberg).

The term *osteitis deformans* was first used by Czerny in 1873 to describe a case of spontaneous curvature of the lower limbs. This was probably a case of "osteomalacia." In 1877 Paget used this term for that group of cases which he described as a separate disease, which disease now commonly bears his name.

**Terminology.**—The various names which have been applied to this disease are: Ostitis deformans; osteitis deformans (Paget); Paget's disease; pseudorachitis senilis; osteomalacia chronica deformans hypertrophica; ostéite ossifiante diffuse (Lancereaux); osteolysis (Lobstein); hypertrophie spongieuse des os; crânioclérose; hyperostose généralisée; ostéite condensante; ostéomalacie hypertrophique benigne; pseudoarachitisme senile; rhumatisme ostéohypertrophique des diaphyses et des os plats; osteomyelite fibreuse; and sclerose osseuse hypertrophique.

**Occurrence.**—Paget's disease is a rather rare condition. Clopton was able in 1906 to collect but 75 cases from the literature. There had been but two cases at the Johns Hopkins Hospital among over 20,000 medical patients. Hurwitz reported the cases from this clinic.<sup>1</sup> Sternberg stated that the cases were about equally divided between the sexes, but Clopton reported that two-thirds of the cases were males.

**Etiology.**—Among the factors suggested are lues, cancer, arteriosclerosis, nervous lesions, muscular work, and gout.

**Heredity.**—In favor of this the following cases may be cited: Chauffard mentions a mother and daughter, and Berger a mother and son similarly affected (Lannelongue<sup>2</sup>). Smith<sup>3</sup> mentions a father, seventy-four years old, who then had had Paget's disease for thirty-five years, and his son forty-two years of age, with Paget's disease then of three years' standing. In each of these cases the disease began when the patient was thirty-nine years of age.

**Pathology.**—This disease affects especially the bones in the long axis of the body and in the following order of frequency: skull, tibiae, femora, pelvis, spine, clavicles, ribs, and the radii. Often, but not always, the long bones are symmetrically affected, but for a long time one or only

<sup>1</sup> *Johns Hopkin's Hosp. Bull.*, 1913, xxiv, 263.

<sup>2</sup> *Bull. de l'acad. de méd.*, 1903, xlix, 299, Discussion.

<sup>3</sup> *Trans. Med. Soc.*, London, 1905, xxvii, 324.



a few bones may be involved. The bones of the face, hands, and feet are least often affected.

The skull is often from one-half to three-quarters of an inch thick, the thickness due for the most part to new bone formation on the outer lamella. Early in the disease the new bone is porous, vascular, and so soft that it can be cut with a knife, but it hardens progressively until in ten or more years it may be like ivory; or it may become loose and friable. There is also some increase in the thickness of the inner table of the skull. The furrows for the vessels, especially that for the middle cerebral artery, seem deepened. There is obliteration of many or all of the sutures. The number of nutrient foramina is greatly increased. Some emissary veins disappear. The horizontal circumference of the skull is increased, *e. g.*, in one of Paget's cases it was 71 cm. and in Stilling's case 64 cm. The bones of the face are as a rule little changed, with the exception of the superciliary ridges which are hypertrophied. In some cases, however, the bones of the face are much affected, a condition called by some hyperostosis cranii, supposed by Paget to be a distinct disease.

"The shafts of all the long bones are white, nodular and massive, and present an appearance of rugged strength and hardness which causes them at a distance to look as though hewn out of stone. The nodules are in the main smooth and rounded, but in parts, as, for instance, the upper end of the humerus, are craggy and irregular. The nutrient foramina are increased in size and number" (Waterhouse).<sup>1</sup> The deposits of new bone follow in general the normal ridges and projections, the deformities, therefore, suggesting exaggerations of the normal shape of the bones.

The long bones of the legs are especially affected. The femur is much thickened and bent, its convexity outward. The girth of this bone at its middle in Waterhouse's patient was five and three-quarter inches. The arrangement of the cancellous tissue of the femur is destroyed. The tibiae are usually and often markedly affected. They are huge and very much bent, their convexities forward, but in some cases and even to an extreme degree the convexity is outward. The tibiae are thickened especially along their anterior margins. The fibulae also are much thickened and bent. The tibiae and fibulae may be connected for some distance by a dense bridge of bone, suggesting the presence of a preëxisting periostitis.

The bones of the upper extremities are much less affected, and yet the changes in them may be very marked. Indeed, in a few cases the disease begins here. The humerus may weigh more than twice as much as a healthy bone of the same length and the girth of this bone at its middle may be four and one-quarter inches instead of two and one-half inches as normal (Waterhouse).

The spine shows a marked kyphosis and often partial ankylosis. The bones of the pelvis if changed at all are only somewhat thickened, though the appearance of the patient usually suggests that the pelvis is much broadened. A heart-shaped pelvis similar to that in osteomalacia has been described.

<sup>1</sup> *Lancet*, 1907, i, 1215.

These bone changes have been carefully studied, and yet little has been added to the description of Paget. Stilling describes the disease as a chronic inflammation, a rarefying osteitis, which begins beneath the periosteum and involves gradually the centre of the bone, with the formation of new Howship's lacunæ, Haversian spaces, and perforating canals. In addition there is also new bone formation, both subperiosteal and myelogenous. The latter process gradually gains on the former and so the bones thicken. The new bone is at first soft and yielding, and bends easily on pressure. The calcification of the new bone may proceed irregularly, when retarded leaving cysts full of cloudy contents, when accelerated producing true bone tumors (Clopton).<sup>1</sup> Von Recklinghausen thought that the first step was an osteomalacia resulting in a marked reduction in the cortex and thus allowing the bones to bend. This was followed by an inflammatory process in the malacic areas, characterized by the formation of fibrous tissue (Clopton). Steel pictured the process as follows: first, the absorption of bone with the enlargement of the Haversian canals; then, the formation of new bone which runs diffusely through the old portions. This new bone remains uncalcified and is, in turn, absorbed. The medullary substance is converted into vascular connective tissue. Lunn thought that constitutional conditions caused atrophy of much of the skeleton. This led to deformity of those bones most used and that to a compensatory hypertrophy.

The result of these changes is a very variable picture. "The enlarged bones are abnormally dense or rarefied. The surface is smooth or rough, nodular or protuberant; the cortex thickened or thinned, spongy or eburnated; the cancellated structure sclerosed or coarsely trabeculated; the narrow spaces obliterated or transformed into cavities of various size. The central canal of the long bones is narrowed or dilated even to disappearance; and the deformity of these bones is further increased by various degrees of abnormal curvature. There is no uniformity in the distribution of these alterations, and a single bone may be evidence of the decalcification, absorption, the formation of osteoid tissue and its calcification, which are the processes concerned in the production of the gross changes. Multiplicity of the bones affected is the constant characteristic." (Fitz.)<sup>2</sup>

It was at first supposed that the abnormal curves in the bones could be explained as the result of tension, and that the new bone formation was a protective compensatory process. But this can hardly be true. Von Recklinghausen pointed out that the hypertrophy was more than compensation would demand and that the distribution of the new bone did not suggest compensation. For instance, the new bone of the tibia is on the convexity, not the concavity, of the twisted bone, while the new bone of the skull could not furnish any mechanical advantage. He suggested that the inflammation which produces the new bone might be due to trauma and thermal influences. These enlarged bones are easily fractured, which accident may lead to the diagnosis.

<sup>1</sup> *Interstate Med. Jour.*, 1906, xiii, 223.

<sup>2</sup> *Trans. Assoc. Am. Phys.*, 1902, xvii, 398.

General arteriosclerosis is a practically constant lesion in cases of Paget's disease. This sclerosis while usually extreme need not be uniform. Bécélère suggested that the bony changes were due to extreme sclerosis of the nutrient arteries of the affected bones. Hypertrophy of the heart is an almost constant feature. Valvular lesions, especially of the mitral valve are common, and seem the result of atheromatous changes rather than of endocarditis.

Joint changes are rare, but in one case both knees showed the lesions of arthritis deformans.

The central nervous system has been carefully studied in the hope of finding the essential lesion of this disease, but while many lesions have been described they can best be explained as the result of arteriosclerosis.

Tumors are not infrequent in cases of Paget's disease. Among these are endothelioma (?) of the pleura and radius (Paget); carcinoma of the liver (Goodhard); tumor of the dura mater (Wilks); sarcoma of the tibia (Wherry); enchondroma of the pelvis (Lunn); cancer of the stomach (Moizard and Bourges), etc. A few cases of Paget's disease have later developed osteosarcoma, especially of the giant-cell variety.

The nature of this disease is unknown. Some have considered it merely an extreme grade of "arthritis deformans." Most observers, however, agree with Paget that it is an inflammation of bone of the nature of which little is known. One of the most interesting recent suggestions is that Paget's disease is luetic in origin. This idea was first seriously advocated by Lannelongue<sup>1</sup> in an article entitled "Note on Hereditary Syphilis of Bones in the Newborn (Parrot's Disease) in Children and Youths, in Adults, and in the Aged (Paget's Disease)." According to this writer Paget's disease is a manifestation of hereditary lues in the aged. He based this opinion on the similarity of the lesions. Fournier agreed with Lannelongue, and mentioned the cases of two brothers, the one of whom developed typical hereditary lues and the other Paget's disease. Fournier later speaks of Paget's disease as a paraluetic condition, to be classed with tabes dorsalis, general paresis, etc. Auffret<sup>2</sup> reported a case of Paget's disease in a patient who probably had hereditary lues, and mentioned other cases. A case reported by Dufour, Bertin and Mourot<sup>3</sup> is that of a woman, aged eighty years, in whom the Wassermann blood test was positive and whose severe neuralgic pains were much relieved by neosalvarsan.

**Definition and Clinical Picture.**—"From these five cases, which, although few, are well-marked and in some points uniform, as well as from a recollection of two more, of which I have no notes, I think we may believe that we have to do with a disease of bones of which the following are the most frequent characters: It begins in middle age or later, and is very slow in progress, may continue for many years without influence on the general health, and may give no other trouble than those which are due to the changes of shape, size and direction of the diseased bones. Even when the skull is hugely thickened, and all its bones exceedingly altered in structure, the mind remains unaffected.

<sup>1</sup> *Bull. de l'acad. de méd.*, 1903, 3s., xlix, 299.

<sup>2</sup> *Rev. d'Orthopédie*, November, 1905.

<sup>3</sup> *Deut. med. Wchnschr.*, 1913, xxxix, 976.

"The disease affects most frequently the long bones of the lower extremities and the skull, and is usually symmetrical. The bones enlarge and soften, and those bearing weight yield and become unnaturally curved and misshapen. The spine, whether by yielding to the weight of the overgrown skull or by change in its own structures, may sink and seem to shorten with greatly increased dorsal and lumbar curves; the pelvis may become wide; the neck of the femur may become nearly horizontal, but the limbs, however misshapen, remain strong and fit to support the trunk.

"In its earlier periods, and sometimes through all its course, the disease is attended with pains in the affected bones, pains widely various in severity, and variously described as rheumatic, gouty, or neuralgic, not especially nocturnal or periodical. It is not attended with fever. No characteristic conditions of urine or feces have been found in it. It is not associated with syphilis or any other known constituent disease, unless it be cancer.

"The bones examined after death show the consequences of an inflammation, affecting in the skull the whole thickness, in the long bones chiefly the compact structure of their walls, and not only the walls of their shafts, but in a very characteristic manner, those of their articular surfaces.

"The changes of structure produced in the earliest periods of the disease have not yet been observed, but it may certainly be believed that they are inflammatory, for the softening is associated with enlargement and with excessive production of imperfectly developed structures, and with increased blood supply. Whether inflammation in any degree continues to the last, or whether after many years of progress any reparative changes ensue, after the matter of a so-called consecutive hardening, is uncertain.

"Holding, then, the disease to be an inflammation of bones, I would suggest that, for brief reference, and for the present, it may be called after its most striking character osteitis deformans. A better name may be given when more is known of it" (Paget<sup>1</sup>).

Five years later Paget<sup>2</sup> reported additional cases and drew the clinical picture more graphically as follows: "It usually affects many bones, most frequently the long bones of the lower extremities, the clavicles, and the vault of the skull. The affected bones become large and heavy, but with such weakening of their structure that those which have to carry weight or to bear much muscular traction become unnaturally curved and misshapen. The disease is very slowly progressive and is felt only in pain, like that of rheumatism or neuralgia, in the affected limbs and in increased heat at the tibiæ. But neither the pain nor the heat are constant, nor do they continue during the whole progress of the disease; and pain has not been observed in the head even in the cases in which the skull was very thickened. There is not any clear evidence of general disturbance of health. In all cases traced to the end of life, death has ensued through some coincident, not evidently associating, disease,

<sup>1</sup> *Trans. Path. Soc.*, London, 1877.

<sup>2</sup> *Med. Chir. Trans.*, London, 1882, lxx, 225.

which has been aggravated by the condition of the bones only in so far as they may have diminished the range of breathing and the general muscular activity.

"At present, with the exception of the seventh case, this disease has been observed as beginning only in persons over forty years old, and has appeared in no usual relation, whether by inheritance or coincidence, with any other disease except gout. In all of the cases I have seen the general appearance, postures, and the movements of the patients have been so alike that these alone might often suffice for the diagnosis of the disease. The most characteristic are the loss of height, indicated by the low position of the hands when the arms are hanging down; the low stooping, with very round shoulders and the head far forward, and with the chin raised as if to clear the upper edge of the sternum; the chest sunken toward the pelvis, the abdomen pendulous; the curved lower limbs, held apart and usually with one advanced in front of the other, and both with knees slightly bent; the ankles overhung by the legs, and the toes turned out. The enlarged cranium, square looking or bossed, may add distinctiveness to these characters, and they are completed in the slow and awkward gait of the patients and in the shallow costal breathing compensated by wide movements of the diaphragm and abdominal wall, and in deep breathing by the uplifted shoulders. I have seen no case in which these characters are imitated except in those of ankylosis of the vertebræ and ribs, and which have been described by Dr. Allen Sturge under the name of spondylitis deformans; but these are easily distinguished by the lower limbs being naturally straight and the clavicles and skull unchanged."

**Symptoms**—The disease, as a rule, begins late in life, the average age of onset being about fifty years. The youngest case reported began at twenty-one years, while the oldest began at eighty-two years of age. The maximum age, however, is difficult to determine since the disease may have existed for years before any objective signs appeared.

The disease begins insidiously, usually with rheumatoid pains in the legs. The patient seldom notes the developing deformity. He may notice that he has to buy larger and larger hats, and he often complains that his legs are getting less and less serviceable, are becoming weak and awkward, and that he tires easily. But his friends will note that he is growing shorter, that his legs are getting more and more bowed, the back bent, the arms relatively longer, and his whole appearance more ape-like. The changes usually begin in one tibia, then extend to the other, and then in irregular sequence the other bones. The bowing of the legs is both anterior and lateral. The lateral bowing may change a condition of knock-knees to one of bow-legs, and the result suggests poorly treated fractures (Waterhouse's case). In some the trouble begins in the skull. The disease is usually progressive, affecting the bones in irregular sequence, although it may remain limited to one bone for years as in one of the writer's cases in which for years the left femur alone was affected. The disease seems to progress until death, which, as a rule, is due to an intercurrent infection, to cancer, or to accident, for Paget's disease itself does not seem to shorten life.

The clinical picture given by Paget and quoted above is so graphic that we will merely add the opinion of many that leontiasis ossea is a form of this disease. If this is true, then in Paget's disease, in addition to the above mentioned symptoms, the changes in the bones of the face may be marked, the cranial nerves much affected, the headache severe, and there may be mental disturbances.

The patient's height may shorten one foot, owing to the kyphosis of the spine and the bowing of the legs. In one of the Johns Hopkins Hospital patients whose height on admission was four feet six and one-quarter inches the shortening seems to have been more than one foot. Pain in the legs, especially below the knees is the commonest symptom of onset. These pains may precede by several years the visible changes in the skeleton. The pains are sometimes "in the arms and legs and around the hips, more in the muscles and sinews than in the bones and joints." They may be worse at night, and often seem to depend on changes in the weather. In some cases these pains are entirely absent, while others, especially those with marked spinal curvature, have pain only on motion. So much do cases vary as regards pains that Joucherey separated a "painful" and a "painless" variety of Paget's disease. The latter cases are less marked and develop more slowly. Fractures of the involved bones are not infrequent.

As the disease progresses the thorax becomes narrow and the ribs so immovable that respiration finally may be purely diaphragmatic. Other symptoms than those mentioned above are unusual. The patients feel "well," for Paget's disease seems to have little influence on the general health. Some patients complain of bronchitis, which is not remarkable considering the rigidity of the chest wall, while cardiac disturbances, valvular and myocardial, are not rare, which is easily explained by the arteriosclerosis: Melanoderma was an interesting feature of Hudela and Heitz's case.

Emphasis has been laid on the claim that in Paget's disease the mind is unaffected, and yet in Fitz's case the "mild stupor, depression and delusions" were sufficient to necessitate confinement in an asylum.

**Diagnosis.**—A fully developed, well-marked case of Paget's disease can usually be recognized at a glance, but an early diagnosis is often difficult. These patients may be treated for years for "muscular rheumatism," "chronic rheumatism," neuralgia, sciatica, senile marasmus, arteriosclerosis, etc. The early cases with involvement of but one bone are particularly difficult of diagnosis, for some have believed that "multiplicity of the bones affected is the constant characteristic." Nevertheless the disease may be for years limited to one bone and since the disease may begin at eighty years of age or over, many patients must die before the second bone becomes involved. This difficulty of diagnosis should be emphasized, since some studied cases may not have been Paget's disease (*e. g.*, the value of the findings in the case studied by Apert, Bornait, and Legueule<sup>1</sup> depends entirely on the correctness of their diagnosis). Many pathologists believe that the bone changes are not specific, others that the

<sup>1</sup> *Bull. et mém. de la Soc. méd. des Hôp. de Paris*, March 14, 1907, p. 235.

diagnosis can be made by the microscopic study of the bone lesions if the pathologist has several bones at his disposal. If, then, a careful pathological study may not be conclusive, how difficult must be the clinical diagnosis. Lues would be a natural diagnosis when one tibia only is affected. When the disease is more fully developed one must exclude osteomalacia, spondylitis deformans, acromegaly, hypertrophic pulmonary osteoarthropathy, and neoplasm. The differential diagnosis between Paget's disease and diffuse or tubercular hyperostosis will depend on one's definitions.

The relationship between *osteomalacia* and Paget's disease may be closer than is generally believed. There are two forms of osteomalacia which have a fair claim to recognition as clinical entities: the osteomalacia of pregnancy especially and senile osteomalacia. The points which osteomalacia and Paget's disease have in common are, kyphosis, the shortening of stature, the bending of the limbs, and the rheumatoid pains. But in osteomalacia there is no hypertrophy of the bones, the involvement is symmetrical, the deformities begin in the skeleton of the trunk and later involve the extremities, the skull is affected only in the severest cases and the pains are much less pronounced. In puerperal osteomalacia the bone changes begin in the pelvis and lumbar spine. In senile osteomalacia pain is prominent, and these patients are bed-ridden early.

*Hyperostosis cranii* is a disease described as involving chiefly the skull. The formation of hyperostoses, the increased intercranial pressure, and cerebral nerve paralysis are its prominent features. Sternberg admitted that there are border-line cases between this and Paget's disease, and now it is believed that they are all one disease.

In *hypertrophic pulmonary osteo-arthritis* the long bones are not bent, the ends of certain bones only are involved, the fingers are much affected, and the cranial bones are not involved.

Cases of *spondylitis deformans* with involvement of the spine and large joints may resemble early cases of Paget's disease, but the entire absence of changes in the long bones is a fundamental difference.

*Acromegaly* bears only a superficial resemblance to Paget's disease. In both there is enlargement of the head, kyphosis, and thickening of the long bones. But in acromegaly the bony lesions are symmetrical. There are characteristic changes in the soft tissues which entirely fail in Paget's disease, and it is the bones of the face which are enlarged rather than those of the cranium.

**Prognosis.**—Paget's disease seems to have almost no influence on the general health, and these patients are, as a rule, remarkably comfortable. If, however, the cases of general hyperostosis are included under this heading then the above statement must be modified, for the prognosis will in the latter group depend on the local pressure (*e. g.*, on the brain) exerted by the bony tumors. Cases of Paget's disease possibly are more susceptible to acute infections than is the normal man of the same age; pulmonary emphysema, chronic bronchitis, etc., would be expected; and arteriosclerosis of a rather severe grade is the rule.

**Treatment.**—There is as yet no treatment of value. One can give the various antineuralgic remedies for the pains, especially the iodides and

quinine. If it is true that Paget's disease is due to lues one would expect improvement under antiluetic treatment, and in some cases this would seem to be the case. The failure of other cases to improve under antiluetic treatment is not a strong argument against the luetic origin, for late tertiary lesions are little influenced by treatment.

**Tumor-building Osteitis Deformans (v. Recklinghausen).**—**General Hyperostosis of the Skeleton with Cyst-building (Virchow).** **Osteomalacia with Cyst-building (Hirschberg).**—This condition seems to be intermediate between Paget's disease and multiple primary neoplasm of bone. The skeleton is much deformed by multiple hyperostoses, fractures, pathological curves, definite tumors and cysts. In the medullary canals in one case (v. Recklinghausen) were multiple fibrocystomas, in another case at exposed points giant-cell sarcomas with cyst formation. In Virchow's case there were colossal hyperostoses of the skull and hyperostoses and curves of some long bones, but osteoporosis of others. In Hirschberg's case there were multiple sarcomas, with cysts and multiple fractures.

### OSTEOGENESIS IMPERFECTA.

**Terminology.**—Osteogenesis imperfecta; Periosteal dyscrasia; Osteomalacia congenita (Jürgens). Rhachitis annularis; Chronic parenchymatous osteitis (Schmidt); Fragilitas ossium congenita (Klebs); Periosteal aplasia (Klebs); Osteoporosis congenita and osteosclerosis congenita (Paltauf); Osteopsathyrosis; Myeloplastic bone aplasia (Kardamatis, 1913).

These terms have been applied to illustrations of a large group of perhaps quite different diseases seen in fetuses, stillborn infants, and very seldom in the living child, which have in common one feature, imperfect bone formation. Formerly all such cases were diagnosed "fetal rickets." Those cases described by some as "annular rickets" or "multiple intra-uterine fractures" are considered by others to be the fetal form of osteopsathyrosis of adults; those of "osteomalacia congenita" are regarded by Jürgens as the congenital form of osteomalacia of adults. In general, newborn infants with osteogenesis imperfecta are, in marked contrast to those with achondroplasia, thin, very wrinkled and with poor muscular development. Since the body proportions are fairly normal the condition may easily be overlooked.

While in achondroplasia the disease would seem to affect the epiphyseal, not the subperiosteal, bone formation, in osteogenesis imperfecta, on the other hand, the subperiosteal bone formation is affected, and the epiphyseal little, or none at all. In osteogenesis imperfecta the bones are of normal length, but their shafts are either soft or fragile. In some cases the long bones present angular deformities, "false joints," "callus masses," and other lesions which are explained as "healed or partially healed fractures;" in others the bones are so soft and pliable that they can be bent easily without crepitation; in others the diaphysis consists of alternate segments of cartilage and bone, while in still other cases the bones are slender and brittle, some indeed containing only parchment-like lamellæ of osseous tissue. Each rib may be studded with cartilage



knots, and the long bones present a succession of thickenings along their diaphyses. The pelvis may resemble that of osteomalacia. The skulls of these infants are especially interesting. The vault of the cranium is in some a membranous pouch in which lie one (in Stilling's case in the forehead) or many little islands of bone with jagged stellate margins. In other cases the ossification of the cranium is extensive but these bones parchment-like. The bones of the base of the skull and of the face are normal. Skiagrams show the cortex of the bones to be thin, the spongiosa soft and porous, and the presence of cavities and cysts. The cavities are full of a gelatinous material. Kardamatis explained the cysts as due to either compression of the lymph vessels and bloodvessels of the marrow, or to a primary disease of the walls of these vessels, leading to widening of the lymph spaces and bloodvessels.

**Etiology.**—At first it was thought that these fetuses seldom lived. Now we are sure that some cases of osteopsathyrosis in children are illustrations of this disease, and Röntgenology shows it to be far more common than was supposed. Some cases are one of twins, the other normal. Zurhelle<sup>1</sup> reported cases of mother and living child both thus affected, which cases he claimed prove the inheritance of osteogenesis imperfecta and the identity of this condition and osteopsathyrosis.

**Pathology.**—Histologically the cartilaginous ossification is normal, but the subperiosteal is abnormal. The osteoblasts are few and scattered; the osteoclasts are abundant. The result is faulty bone formation and the reduction by absorption of the diaphyseal cortex to thin lamellæ which had been formed.

Since one meets the term intra-uterine fracture so often, a short discussion of this subject may be in place. It sometimes, although rarely, happens that one or more bones of an infant are actually broken, usually during or recently before birth. These genuine fractures are usually single. While genuine fractures of a normal fetus knit rapidly, the so-called fractures under discussion must have knit very slowly, since the lesions thus interpreted are sometimes false joints, the ends of the fragments being atrophied and pointed, or separated by spaces filled by cartilage; other cases have fractures of very fragile bones which consist of parchment-like lamellæ of bony tissue; but in by far the majority of cases the lesions described as fractures are enlargements along the shaft of the bone, suggesting callus formation, or angular deformities, which suggest healed breaks. One may well doubt, however, that many, if indeed any, of these lesions are the results of real fractures. Direct violence could scarcely produce the multiple "fractures" (even 113 in Chaussier's case) which these infants show; these bones thus affected are not always more brittle than normal; the callus and angular deformities are at positions (*e. g.*, near epiphyses) where a mechanical pull or a blow would not be likely to fracture the bone; and, finally, there is a suspicious symmetry in the arrangement of these lesions. The cases with multiple callus formations have long borne the diagnosis "annular rickets," the rings of callus being explained as irregular proliferations of

<sup>1</sup> *Deut. med. Wchschr.*, 1913, xxxix, 1484.

the periosteum. The angular deformities are often, and some think usually, due to ingrowths of periosteum between diaphysis and epiphysis on one side only of the bone, thus inhibiting the growth of the bone on this side. In favor of the view that these lesions are developmental abnormalities rather than fractures is the partial or total absence of lesions of the fibula when the tibia is involved. Some cases of this group seem due to lues.

**Osteitis Fibrosa.**—As osteitis fibrosa has been described a condition characterized by the rarefaction in localized areas of the bone tissue and its substitution by fibrous tissue or by a cyst (*osteitis fibrosa cystica*) even the size of a hen's egg. These areas are often mistaken for sarcomas. Some have followed trauma.

### OSTEOPSATHYROSIS.

Osteopsathyrosis (Lobstein, 1833); *Fragilitas ossium* (Klebs); Lobstein's disease. This rare condition, characterized by abnormal brittleness of the bones, occurs in infancy, childhood, and adult life.

Abnormal fragility of the bones occurs in old age (senile osteoporosis), in cases with atrophy of disuse, in cases of cachexia (especially that due to malignant disease), in various diseases of metabolism as rickets and scurvy, in certain nervous diseases, as locomotor ataxia, general paresis and anterior poliomyelitis, and in general intoxication, as phosphorus poisoning. Among the insane a special form of *fragilitas ossium* occurs, which is due to rarefaction of the bones. The atrophy of the bony tissue in the above mentioned groups of cases may be marked. These conditions are ascribed to poor nutrition and to the frequent traumas which these patients suffer. Localized brittleness of bones may be due to the local osseous lesions of lues, osteosarcoma, the metastases of carcinoma, multiple myelomas, and to echinococcus cysts. In all the above conditions examination will reveal local lesions, gross or microscopic changes in the bones, which easily explain their brittleness.

The term osteopsathyrosis, or Lobstein's disease, etc., is reserved for "idiopathic cases," cases "without a demonstrable (as yet) lesion." It is true that radiographs of the long bones of these cases would suggest that the cortex is rather thin, the shafts slender, and for this reason, perhaps, the epiphyses are apparently enlarged, and yet the lesions would seem scarcely sufficient to explain the fragility. If osteopsathyrosis is osteogenesis imperfecta of viable infants, then the brittleness of the bones is due to the inability of the periosteum to develop a sufficiently thick cortex; others think that it is due to relatively overactive absorption of the cortex. Some think it secondary to changes in the nervous system. Lipschutz thinks the lesion lies in the cancellous tissue, since in his case the fractures involved the extremities of the bones, and the skiagraphs showed that here the trabeculae were abnormally few. Zesos suggested that muscular atrophy is a part of the same condition and that the disease is related to insufficiency of glands of internal secretion. One case is reported to have improved after the transplantation of the thyroid.

These cases would seem to be rare, since so few are on record. The sexes are equally affected. In a rather large proportion the condition is inherited and in still more cases it affects several of the same generation. The general health of these patients would appear to be excellent. The fractures follow very trivial injuries, as a slight blow, muscle pull, a fall, and turning over in bed. Simple chewing may fracture the jaw. The long bones are the ones most frequently broken. The number of fractures which these patients may have during their whole life varies from a few to more than one hundred.

This disease affects especially the period of development. The first fracture may occur during birth, during the first bath, or much later, even in the fourteenth year and in adult life. In most cases the condition ceases at thirty years of age, but in one case it continued until the seventieth year. In some cases the bones would seem earlier to be soft and pliable and later rigid and fragile.

**Symptoms.**—Usually the fractures are rather painless and as a rule they knit quickly, more rarely slowly. The amount of callus thrown out varies much. The patients usually are markedly deformed. These deformities are due for the most part to the union of the fragments in bad positions, and yet some of the deformities develop during a stage when the bones are abnormally soft and pliable. This applies especially to the pelvis. The tibia is often sabre-shaped, wide, and flattened laterally. In one case there was progressive ankylosis of the joints.

**Prognosis.**—Osteopsathyrosis can make a patient wretched for years and then in middle life the bones become stronger. In these cases the prognosis is one of function rather than life; the deformities resulting from union in faulty positions may be a great calamity.

**Treatment.**—The only medication suggested which seems to have been beneficial is phosphorus and transplantation of the thyroid gland. The treatment of the fractures is a surgical problem.

### HYPERTROPHIC PULMONARY OSTEO-ARTHROPATHY.

**Synonyms.**—Toxigenic ossifying osteo-periostitis; osteo-arthritis hypertrophique; secondary hypertrophic osteo-arthritis; clubbed fingers; akro-elephantiasis ossea et mollis.

**Definition.**—The above terms have been applied to conditions characterized by symmetrical enlargement of the ends of the fingers and toes and of the distal ends of certain long bones. These lesions do not constitute independent diseases, but are secondary to diseases of certain organs, especially the lungs, or to a local neuritis.

**Etiology.**—Marie considered that this condition was always secondary to other diseases, usually of the lungs, because, of the 8 cases he studied, 4 certainly and 3 possibly had pulmonary trouble. Wynn<sup>1</sup> in 100 cases, from the literature, found pulmonary trouble in 68; Alexander in 77 of 103 cases (bronchiectasis in 25, chronic pulmonary tuberculosis in 15, empyema in 8, malignant disease of the mediastinum or lung in 8, chronic

<sup>1</sup> *Birmingham Med. Review*, 1904, lv, p. 212.

bronchitis in 28, pneumonia and pleurisy in 2, and a variety of pulmonary diseases in 17. Only two cases, one of which was congenital, did not show a definite antecedent disease.

Alexander, who was very critical in his selection of cases, found that osteo-arthropathy occurs usually with pulmonary troubles, especially those with a septic focus as bronchiectasis. In other cases the pulmonary lesion was chronic pulmonary congestion and chronic passive congestion of the whole body. Marie ascribes the changes to toxins produced in a focus of infection. This theory may explain the majority of the cases, but certainly not those with congenital heart disease and clubbed fingers. Some claim that cyanosis causes tissue proliferation, but there are many patients with extreme and chronic cyanosis whose fingers are not clubbed. Shaw<sup>1</sup> claims that his is the only case of hypertrophic osteo-arthropathy associated with congenital heart disease without any pulmonary lesion.

Other diseases claimed as primary are hypertrophic cirrhosis of the liver and lues. The neuropathic theory also has many adherents. In favor of this are the associated paresthesias, the sensory changes suggesting syringomyelia, epileptiform attacks, and the marked sweating. That clubbing of the fingers may follow a traumatic neuritis is granted, as in Möbius' case of neuritis of the ulnar nerve with extreme clubbing of the two fingers which that nerve supplies. This patient had tuberculosis also. On the other hand in chronic hypertrophic pulmonary osteo-arthropathy the trophic changes in the skin so common in neuritis are not mentioned, and as yet no lesion of the nervous system has been found. Clubbing of the fingers and swelling of the distal end of the forearm and less in the upper arm of the left side has been reported in cases of subclavian aneurism, in aneurism of the arch of the aorta, and in cases with unreduced dislocation of the shoulder. In these cases the clubbing may have been due to pressure on the nerves or to passive congestion. Congestion alone, however, could hardly cause this. The pressure of aneurism on the nerve trunks probably causes a severe neuritis which leads to the hypertrophy.

That *tuberculosis* is the chief cause is a theory just now attracting attention. Thorburg advocated this in 1893.<sup>2</sup> Poncet and his pupils consider it to be benign tuberculous rheumatism and Alamantine<sup>3</sup> proposes to rename the disease "subacute tuberculous osteo-arthritis." They, it would seem, do not claim that Marie's syndrome is tuberculosis of the periosteum or bone, but that it is caused by toxins from a tuberculous focus and so they agree with Marie. The specific lesions of tuberculosis have never been found in the involved bones. Trauma is said to predispose the bone to this condition. Italian writers claim that the germs or toxins which cause the bone lesions do so by first causing polyglandular disturbances. Others would separate the cases without a discoverable primary disease into a separate group with which the congenital cases might be grouped. The term *akro-elephantiasis ossea et mollis* has been proposed for cases in which neither the lungs nor the heart are diseased.

<sup>1</sup> *Clin. Soc. Trans.*, 1907, xl, 259.

<sup>2</sup> *Brit. Méd. Jour.*, 1893, i, 1155.

<sup>3</sup> *Rev. de Chir.*, 1907, xxxv, 992.

Sternberg concludes that, since the etiology is so varied, one must suspect that the cases diagnosed as chronic hypertrophic pulmonary osteo-arthritis are not all the same disease. The periosteal changes and the clubbing of the fingers may not be related. We do see clubbed fingers without periosteal changes in long bones, but not the reverse.

*Race*.—It is interesting that all but 3 of the 117 cases reviewed were of the white races.

*Sex*.—Of the 77 carefully selected cases collected by Alexander, 64 were men and 13 women. Landis states that males are eight times more frequently affected than females.

*Age*.—While all ages are affected, in the majority of cases the onset is in the third and fourth decades. In a few cases the condition seemed congenital, even inherited (Marie).

We know nothing of the conditions immediately determining the development of the disease. Bronchiectasis or chronic pulmonary tuberculosis may continue for years and then suddenly the hypertrophic osteo-arthritis may appear.

**Pathology.**—There are reports of but sixteen complete autopsies. These are well reviewed by Alexander.<sup>1</sup> The lesions of chronic hypertrophic pulmonary osteo-arthritis should be carefully separated from those of the primary trouble. Its characteristic lesion is a symmetrical deposit of new subperiosteal bone on the shafts of the long bones. The bones most frequently affected are the lower ends of the radius and ulna, the metacarpals, and the first two rows of phalanges. More rarely the lower end of the humerus and the upper ends of the radius and ulna are involved. In the forearm the new bone begins abruptly as a thin layer, about four or five inches above the wrist-joint, and forms a sheath covering the lower ends of the ulna and radius as far as the epiphyseal line. The circumference of the shaft is about equally affected. The layer of new bone is generally thickest at the juncture of the shaft with the lower extremity of the bone. In Landis' case there was an osteophyte growing from the olecranon. The carpal bones are not affected, but the shafts of the metacarpals are ensheathed and so appear to be uniformly enlarged. The same is true of the first and second rows of the phalanges, but the change is less marked than on the metacarpals. The terminal phalanges are apparently unaffected. The corresponding bones of the lower limb, the lower ends of the tibia and fibula especially, are affected. The deposit of new bone on these bones is more diffuse, and may extend over the whole shaft, covering also their upper ends. The layer of new bone is thicker on the tibia than elsewhere. Here it may be a quarter of an inch thick. Sometimes the lower end of the femur and more rarely the upper ends of the tibia and fibula also are covered. The tarsal bones are not affected. The metatarsal bones show changes similar to those of the metacarpals, but the phalanges of the toes are less frequently affected than of the fingers. Deposits of new bone have been found on the clavicles, iliac crests, and the patellæ. The symmetrical arrangement of the lesions is remarkable.

<sup>1</sup> *St. Bartholomew's Hosp. Rep.*, 1906, xlii, 41; see also Sternberg, *Nothnagel's System*, 1903, Band vii, Part 2.

The cortex of the bone may show some sclerosis and thickening, with diminution in the size of the medullary cavity. The marrow is said to be embryonic in character, with a tendency to fatty degeneration in its central part (Wynn). In one case at least the cortex of the shafts was softer than normal (Alexander). The periosteum is thick and abnormally vascular over the new bone. The main nutrient canals are large, and many additional vessels enter the compact layer. The skull is little if any affected. Marie reported one case with marked thickening of the upper jaw, but no similar case has since been reported. These bone changes are due to a chronic inflammation which varies enormously in extent and severity. The bone changes are only part, and often a small part, of the process. There is much thickening of the connective tissue covering the affected bones, especially in the neighborhood of the wrists, ankles, and fingers. The clubbing of the fingers is due entirely to changes in the soft tissues of the finger tips. These changes are: increase of fat, an overgrowth of the papillæ (Fowler and Godlee), dilatation of the capillary loops in the nail beds (Freytag). The clubbing disappeared in the finger preserved by Fowler in alcohol, also in the finger which Landis preserved in Kaiserling's fluid. When the x-rays were first used the clubbing was supposed to be due to the irregular spicules of bone radiating from the terminal phalanx, but these are not more common or more prominent in the clubbed than in some normal fingers.

In many cases there is increase of fluid in the joints, especially the wrists, knees, ankles and fingers, due to subacute synovitis. As a rule the cartilages show no changes. There is little lipping, little osteophyte formation, and little eburnation of the joints. The thyroid gland and the pituitary body are normal.

**Symptoms.**—The first symptoms may be pain, or clumsiness in fine movements, stiffness of the joints, or muscular weakness. In some cases the acute stage is rather brief, the condition in a few months passing from its acute to its chronic stage. The bone lesions once developed may progress no further, although the pulmonary condition which it complicates may progress without improvement.

In describing this condition we follow for the most part Marie's original description (see also Alexander, Thayer,<sup>1</sup> Wynn, and Landis). The *hands* appear enormous, even larger than in acromegaly. Their general relative proportions are not, as in acromegaly, fairly normal, but the hands are deformed and resemble somewhat an animal's paw. The fingers especially are affected, particularly the terminal phalanges, and have the shape of a drumstick. Sometimes the greatest swelling is not terminal, and the fingers are spindle-shaped. Marie mentions a patient in whom the terminal phalanx of the middle finger measured 10 cm. in circumference. The fingers often seem somewhat longer than normal. Hyperextension of the terminal phalanx has been noted. The joints of the fingers are sometimes swollen and their movements so limited that the hand cannot be completely closed.

The *nails* are large, even 2.5 cm. broad, and curved both longitudinally

<sup>1</sup> *New York Med. Jour.*, 1896, lxiii, 33.

and transversely. They may overlies their beds laterally and reach the border of the finger. If the nail was previously long, the end of the finger, when viewed in profile, has the shape of a parrot's head. If previously short, the nail becomes round and fits on the finger like a watch crystal. They often resemble the bowl of a spoon. These nails show marked longitudinal striation and are brittle. The root of the nail is elevated and almost fluctuating. The root of the nail of a normal finger is depressed, rather firmly fixed to the periosteum, and when pressed at its root yields very little. The slightest grade of clubbing of the fingers is evidenced by the fact that its root can be depressed on pressure. In more marked cases the entire dorsum of the nail including its root is in a line with the dorsum of the finger. In an extreme case the dorsum of the finger is convex from the last knuckle to the tip of the nail, the root of the nail forming the summit of the convexity. In these cases the root of the nail "floats" when pressed and projects as a ridge when the free end of the nail is pressed. This occurs in no other condition (Wynn). The nails grow with abnormal rapidity.

The tips of the fingers are cyanotic, but there is usually a zone of bright rose color at the periphery of the nails. As a rule all of the fingers are affected, but the thumb, index and middle fingers are often more affected than the other two. Sometimes one or two fingers alone are affected (possibly those which have received more traumatism).

The forearm above the wrist is so much enlarged that from elbow to wrist it may have a uniform circumference. The ulna and radius at the wrist, especially the latter, project prominently.

Changes similar to those in the hand, but usually less in degree, occur in the feet and ankles. The malleoli are quite prominent and the great toe especially is affected. Since the lower end of the femur and the whole of the tibia and fibula are enlarged, the knee-joint stands out prominently and the whole leg may have an elephantine appearance. This hypertrophy may affect the other long bones, especially at their extremities, the humerus and femur, the clavicle, sternum, ribs, and the iliac crests. With improvement in the general condition the swelling may diminish.

The lesions of the long bones are usually painful, both spontaneously and on pressure. In some cases the pains are persistent, but more often they are severe at certain times, particularly in the evening. They may be sharp and stabbing and last from a few minutes to half an hour. These pains are increased on motion and on exposure to cold, and relieved by rest and warmth. They lessen or disappear when osteosclerosis begins. Certain bones which are not swollen may be painful on pressure.

In some cases the symptoms of arthritis are conspicuous, but the joints usually appear more involved than is actually the case. They are swollen and painful, but not red, and there is increase in the synovial fluid. The joints nearest the affected bones are most involved. The wrists and ankles are the ones most affected, less often the elbows, the shoulders, and the small joints of the hands and feet. The hips usually escape. The temporomaxillary and sternoclavicular joints practically never are involved.

The swelling of the bones and joints and the tenderness on pressure are the only evidences of inflammation. The joints are never red nor hot, nor do they suppurate. There is very little stiffness on motion. But even these signs of inflammation are seen only in rapidly developing cases or during an acute exacerbation of an otherwise painless case. It is reported that in some cases complicating bronchiectasis acute attacks occur which last for two or three days, with pain and swelling of the joints, which attacks are coincident with a temporary diminution in the amount of sputum (Alexander).

Kyphosis, especially of the lower thoracic spine, occurs but this appears to be accidental. Atrophy of the muscles of the limbs is sometimes a marked feature and because of it the swelling of the bones is made more conspicuous. This atrophy, however, affects the muscles of the proximal segments and is not symmetrical.

In one case there was a noticeable deformity of the upper jaw. Clubbing of the tip of the nose has been mentioned. There is often true oedema of the hands and feet. Excessive sweating is a very common feature. This may be general, or limited to the feet, ankles and hands. Skin eruptions are common in these cases; among these are ichthyosis, eczema, pigmentation, erythema, hypertrichosis, etc. The urine is, as a rule, negative, although an accompanying polyuria has been described.

**Clinical Types.**—Sternberg separated three types, which may be simply three grades: 1. In the mildest type is found only clubbing of the fingers which occurs in patients with chronic purulent diseases of the lung and in those with congenital heart disease. In empyema and acute tuberculous pneumonia the clubbing can develop rapidly, even in a few weeks. After the cure of the empyema by operation the clubbing can disappear almost as suddenly as it developed. As a rule clubbed fingers are not painful. The attempts to distinguish between simple clubbing of the fingers such as occurs in heart disease and that seen in hypertrophic osteo-arthropathy have thus far been unsuccessful.

2. This group includes cases with clubbed fingers and also a painful thickening of the long bones, especially those of the forearm and lower legs. This, Bamberger's type, occurs with the same diseases as clubbed fingers, and occurs also in chronic jaundice.

3. The third is "Marie's type," or osteo-arthritis hypertrophica. While in the first two groups the lesions are merely incidents in the course of a disease, in this third group the bone changes are the most conspicuous features and amount to actual deformities. The clubbing of the fingers is at a maximum. The hands are huge, hideous, and "paw-like." The forearms are diffusely thickened. The feet are of giant size, especially the toes and the malleoli. In this form the pelvis, sternum, ribs, and the clavicles are often thickened, and there may be kyphosis of the spine. The symptoms of arthritis are most marked and the patient uses his extremities with painful difficulty. In this group the primary diseases may be in the background, or even remain undiscovered. Among them are: Sarcoma of the lung, multiple sarcoma with pleural exudate, pulmonary tuberculosis, empyema, carcinoma of the lung, bronchitis, influenza, bronchiectasis, etc.



Alexander classifies the cases of osteo-arthritis as acute and chronic. In the acute cases the joint symptoms are prominent features.

Since hypertrophic osteo-arthritis is a secondary condition, as might be expected, a large number of atypical cases occur. Of the 103 cases which Alexander collected, 26 were not typical. Bamberger's case seems to be even unique. All the long bones of the patient were more or less completely covered with a stratified layer of bone, porous in some places, harder in others, and thickest over the normal ridges and spurs to which muscles are attached. Here and there small areas of unaltered bone were left. Métin and Guillon<sup>1</sup> reported a case which, because of the shape of the hand, and especially of the thumbs, they described as "hypertrophic pulmonary osteo-arthritis of the acromegalic type." There were no changes in the face or head.

**Diagnosis.**—In every case of chronic pulmonary disease the appearance of pain and swelling near the joints should lead one to examine for hypertrophic osteo-arthritis. The x-ray examination is very important. In a well-marked case the shadow of the sheath of new bone is clearly differentiated from that of the compact bone. The outlines of the bones are usually smooth. The metacarpal bones are among the first to be affected. The terminal phalanges of even markedly clubbed fingers are practically normal.

Clubbed fingers should not be confused with Heberden's nodes. These are small exostoses on the dorsal side of the proximal ends of the terminal phalanges. They may be of even pea-size, and are sometimes painful. They are often associated with arthritis deformans. These phalangeal joints are often ankylosed, sometimes at an angle. Heberden's nodes are due to a thickening of the tubercles to which the tendons of the extensor muscles are attached. They are not the same as Bouchard's nodes, which develop at the joints between the proximal and middle phalanges, and also at a point between the metacarpal and proximal phalanx of the thumb. They occur especially in patients with gastrectasis. Meynet's nodes are movable nodules in the joint capsule, sinews or sinew sheaths which occur especially in chronic arthritis. Multiple enchondromas are usually symmetrical. They occur on the hands and feet and on the terminal phalanges.

Bamberger's type of hypertrophic osteo-arthritis is to be distinguished from chronic arthritis deformans. This usually is easy unless both diseases are present in the same patient. Marie's type should be distinguished with ease from acromegaly, although at first these two diseases were grouped together.

**Prognosis.**—This seems to rest entirely with the primary condition, for the osteo-arthritis seems to influence neither the general condition of the patient nor the course of the primary disease. Its onset is usually fairly acute and then this condition remains stationary; but what in the primary disease determines either the development of the osteo-arthritis, the length of the acute attack, its severity or its distribution is unknown. Since the condition is very rare, while the infections which it complicates are common enough, and since its course would seem to

<sup>1</sup> *Bull. méd.*, 1907, xxi.

have no observable relation to that of the disease considered primary, one must be skeptical as to an immediate relationship between the two and suspect that some intermediate condition, as a toxic or traumatic neuritis, determines the bone lesions and controls their prognosis.

**Treatment.**—It is of prime importance to treat the primary disease, and marked improvement in the bony condition has followed successful treatment. One can do much to relieve the pains in the bones and joints. For this warm applications seem efficacious. Demons and Binant have used subcutaneous injection of lung extract with doubtful success.

### GENERAL HYPEROSTOSIS.

Under the term "general hyperostosis" are grouped cases in which the disease affects the entire skeleton; "general hyperostosis of the skull," those in which the skull only is involved. Some doubt that the lesions are ever entirely limited to the skull, but point out that the other bones are often so slightly diseased that these lesions would be easily overlooked. Many of the reports are descriptions of skulls in museums, the rest of the skeletons not preserved; only recently have clinical reports appeared.

**General Hyperostosis of the Skull, Leontiasis Ossea (Virchow), Cephalomegalia, Megalocephlia, Ostitis Fibrosa Hyperostotica Cranii et Faciei.**—These names have been given to a rare condition characterized by diffuse hypertrophy of the bones of the skull. All the bones of the face and cranium are affected, but in varying degree. The result is a huge skull weighing even 4000 grams, and much deformed. The surface of the bones is rough, yet there are no large exostoses. The air sinuses of the face may disappear, the orbits are encroached upon, the sutures disappear, the foramina, especially those for nerves, are narrowed, some of the grooves for the vessels are widened, some narrowed, even obliterating the vessels. The thickened bones may be as hard as ivory or the cancellous tissue may persist.

The interesting question is the relation of this condition to general hyperostosis and of both to osteitis deformans. Paget considered the diseases distinct. Sternberg<sup>1</sup> admitted that there are border-line cases. Lately the opinion has gained ground that hyperostosis cranii is not a disease entity, but a condition common to perhaps several diseases. One<sup>2</sup> of Sternberg's 12 cases turned out later to be a case of syringomyelia; at least one other (Ede's case<sup>3</sup>), of Paget's disease, and still another of lues (Putnam). Prince<sup>4</sup> said: "We have no sure ground for differentiating hyperostosis cranii from osteitis deformans. Both are probably trophic disturbances, perhaps allied, and possibly only different manifestations of one and the same disease." Of course, the differences between these two diseases cannot be overlooked. The cases described as hyperostosis cranii have, for the most part, developed at an earlier age than do the majority of cases of Paget's disease. In the former the lesions on the skull are much more marked, and the symptoms from them much more

<sup>1</sup> Thesis, Paris, 1907.

<sup>3</sup> *Am. Jour. Med. Sc.*, 1896.

<sup>2</sup> Hale White, *British Med. Jour.*, 1896, p. 1377.

<sup>4</sup> *Trans. Assoc. Am. Phys* 1902, xvii, 382.

pronounced than in typical Paget's disease. But these differences are by no means fundamental and may depend on the localization of the lesions. According to a later view the condition should be named "*ostitis fibrosa hyperostotica cranii et faciei*" and regarded as an inflammation caused by trauma, bacterial infection or lues.

**Clinical Course and Symptoms.**—Of the 21 cases collected by Prince, some began in late childhood or puberty, only 10 after thirty years of age, and of these only 4 after forty years. The disease begins insidiously with headaches and a general increase in the volume of the skull and a marked bulging of the forehead. The result is a huge, misshapen head. The headaches are sometimes continuous, sometimes paroxysmal, and are accompanied by symptoms of compression of the nerves in the foramina; neuralgia, blindness, deafness, bilateral facial paralysis, and disturbances in chewing, swallowing, and breathing, with paralyses. The circulation of the head changes considerably, owing to stenosis or obliteration of the vessels in the foramina. Some subcutaneous veins may be very much distended. Stenosis of the canalis caroticus may cause a loud roaring in the ears, diminished by pressure on the carotid artery (Putnam). Exophthalmos is often present. Insomnia and mental apathy develop later; mental disturbances are common; epileptiform convulsions and finally paralysis of the extremities "complete the sad picture." Death follows paralysis of the muscles involved in mastication and swallowing, or, as often happens in cases of chronic intracerebral compression, it may come suddenly with convulsions. The duration is given as from twenty to thirty years.

**Treatment.**—The inflamed areas resulting in hypertrophy should be excised early; later cerebral decompression may be necessary.

**Nodular Hyperostosis of the Skull.**—This form of leontiasis ossea differs from the preceding, or diffuse form, in that the hyperostoses are more definitely tumors on a more normal bone. These tumors may be flat or mound-like, and are of various sizes and shapes, but are seldom prominent and warty. The underlying bone is not perfectly normal, but is the seat of a diffuse hypertrophy of varying degree. The results are hideous deformities. The most common situation for these tumors is the cranium. Situated here they may grow inward, exerting pressure on the brain. In some cases the tumors at first are limited to the lower jaw, but later develop on other bones as well. One form, in which the tumors are for a long time at least limited to the jaw bones and are symmetrically placed, has been described as an independent disease under the name "hyperostosis maxillarum."

So often is this condition combined with giant growth that Sternberg suspects some relation between these two conditions. A somewhat similar though much less marked development of hyperostosis is seen in idiots and the insane.

**Pathology.**—These hyperostoses cannot be sharply differentiated from exostoses and osteomas. They cannot always be differentiated from sarcomas, even on microscopic examination. Gummas must be excluded.

**Symptoms.**—The tumors develop slowly and cause symptoms which depend on the organ pressed against. The most important group of symp-

toms are those resulting from pressure on the brain, on the eyeball, on the respiratory passages, etc.

**Prognosis.**—This depends on the situation of the tumors. When their pressure does not interfere with the function of any important organ the disease may not appreciably influence life. The deformities may be hideous, and while the effects of pressure from such slowly-growing tumors may be much less marked than from the more rapidly-growing ones, yet these patients may suffer severely.

**Treatment.**—This must be local, since the cause of the condition is not yet known. It is always well to try antiluetic treatment in the hope that this may benefit the patient, but, this failing, surgical removal of those tumors whose presence is a menace to life or a detriment to important functions is indicated.

Daireoff<sup>1</sup> thinks that in addition to the above forms there is another group of the systematic osteopathies which he describes as *deforming hyperostosis*. This affects symmetrically the inferior epiphyses of the bones of the legs and forearms and the bones of the feet and hands. The result is elephantiasis-like extremities. The osseous hypertrophy develops slowly, and is preceded by a period of functional trouble. It seems a trophic disturbance.

### MICROCEPHALIA.

The most important features of microcephalia are retardation in the development of the cerebral cortex and mental reduction. Marked grades of microcephalia are rare.

The cases may be divided into two groups, those the result of antenatal disease, and those due to developmental deficiency. The latter group of the "idiopathic" cases are ascribed to "inhibition" of growth of the brain due to pathological "feebleness" of the embryonic tissue from which the cerebral hemisphere will develop. This is the "atavistic" group or the "primary" group of Giacomini. While these cases may be relatively numerous, as yet but one case has been carefully studied pathologically and every trace of antenatal disease excluded.<sup>2</sup> This case was that of a woman, aged forty-nine years, whose brain weighed but 370 grams. The convolutions were abnormal in their arrangement. The woman had also a parenchymatous goitre. He states that this woman's mentality was much better than one would expect.

If we accept as correct the diagnoses of the cases reported as primary, the following is their clinical picture: The cranium is small, even but twelve inches in circumference (this patient was a child twelve years old, Shuttleworth), yet often fairly well-shaped and sutures normal; the face always relatively and sometimes absolutely large; the brain small, with primitive arrangements of the convolutions. The supposition is that the brain does not grow and therefore the skull also does not. There are no indications of intracranial pressure and no signs of brain lesions, such as motor disturbances, convulsions, etc. The reflexes are usually normal. Coördination of all the voluntary movements is perfect, although

<sup>1</sup> *Thesis, Paris, 1900.*

<sup>2</sup> Hilty, *Arb. aus. d. Hirnanat. Inst. Zurich, 1907, ii, 205.*

the movements sometimes are remarkably quick, being described as ape-like.<sup>1</sup> The intellectual development varies much. Walking is delayed, *e. g.*, to the fourth year. Some of these patients never try to talk, and yet they may be able to recognize their friends.

The microcephalia of the cases of the second group is ascribed to antenatal intracranial disease. What these diseases may be is a disputed question. They would seem to resemble in part those producing hydrocephalus, in part those producing porencephalus. The head is small and very narrow laterally, hence resembling a tent; the forehead is very narrow, pointed above, and receding. The sutures have been described as prematurely united, and yet of the 14 skulls studied by Glüh<sup>2</sup> in not one were all ossified. These cases present motor disturbances, spastic paraplegia, anesthetics, and usually exaggerated reflexes. Among these cases there may be a few in which premature union of the sutures inhibits the growth of the brain. With this idea in view surgeons have attempted to relieve the condition by operation, but with very doubtful results.

### FACIAL HEMIATROPHY.

**Synonyms.**—Neurotic progressive facial atrophy; Hemiatrophia faciei progressiva (Parry, 1825).

In this disease there is a slow, progressive wasting of the skin first (some say of the fat), then of the subcutaneous tissue, later of the bones and last and least of the facial muscles. Usually one side alone of the face is affected, seldom both. Not over 200 cases are on record. The majority of the cases are women. The left side of the face is more often affected than is the right. The disease begins, in a majority of cases, during childhood and youth, and there is good evidence that in some cases the condition has a congenital *anlage*, *i. e.*, in Harbitz's case the disease on the left side of the face was accompanied by acromegaly and congenital absence of the left kidney. As immediate causes have been mentioned exposure to cold, insignificant traumas, acute infectious diseases and infections from the tonsils. Believed formerly by some to be due to disease of the local tissues, it is now granted by most to be due chiefly to disease of the trigeminus of the affected side, although other nerves in some cases are involved, as branches of the facial nerve, the cervical sympathetic nerves, and in one case the left radial nerve, causing atrophy of the muscles and skin which it supplies. In the only well-reported autopsy was found the terminal stage of an interstitial neuritis from origin to periphery of all branches of the fifth nerve, especially the superior maxillary branch.

**Symptoms.**—The onset of the atrophy, usually insidious and without subjective symptoms, is sometimes preceded by sensory and motor symptoms in the region of the fifth nerve of the side later affected; neuralgic pains, hyperesthesias, paresthesias, spasm of the masseter muscle and even epileptiform convulsions. The first objective evidence

<sup>1</sup> Jones, *Brit. Jour. Child. Dis.*, 1905, ii, 214.

<sup>2</sup> *Deut. med. Wchschr.*, 1912, xxxviii, 1440.

is thinning of the skin, usually over a wide area but sometimes at one or more small patches of leukoderma which later coalesce. The area of atrophy increases; the subcutaneous fat gradually disappears, and soon the skin seems attached directly to the bone. The bones, especially the superior maxillary, are early affected, and the muscles last and least. The hair of the scalp, the eyebrows, eyelashes, and the beard over the affected area may fall out, or turn white, diffusely or in patches. The activity of the sebaceous glands ceases, that of the sweat glands may continue almost normally. The skin becomes rough and hard, and resembles somewhat that of scleroderma. The patient complains of twitching of the skin and a drawn feeling. The skin sensations are little affected, the skin reflexes remain normal, although the vasodilator reflex on that side may be lost. The eyeball sinks because of the loss of orbital fat. The muscles of mastication become weak early but the facial muscles are little involved. Their reactions on Faradic and galvanic stimulation are prompt. The half of the tongue and soft palate corresponding to the facial involvement are sometimes markedly atrophied. The bones of the face waste, and so their normal eminences become less prominent. The alveolar process especially suffers and the teeth drop out. The features are gradually drawn to the affected side. The result is that the affected side of the face becomes much smaller than the other, and the two sides seem like the faces of different persons. The lesion stops abruptly at the mid-line. The course is slow and progressive until the atrophy reaches a certain grade, after which time the condition may for a long time remain unchanged. Spontaneous improvement may occur.

**Diagnosis.**—In this, one must exclude scleroderma, the atrophy following nuclear lesions and paralysis of the cervical sympathetic nerve, that following anterior poliomyelitis, and the facial atrophy seen in hemiplegia. The face is asymmetrical in congenital wry-necks. The normal side looks atrophied in acquired facial hemi-hypertrophy.

**Prognosis.**—This condition does not seem to influence appreciably the general health. When once it begins there is nothing which seems to stay its course, and the periods of improvement cannot be attributed, so far as we know, to any therapeutic measure.

**Treatment.**—There is no treatment which affects the course but the appearance has been much improved by paraffin injections.

# INDEX TO VOLUME V.

## A

ABDOMINAL reflexes, anomalies of, 78  
 Abducens nerves, anatomy of, 483  
 Abscess of brain, 378  
     of spinal cord, 254  
 Abscesses, metastatic, diagnosis of, from  
     primary suppurative myositis, 874  
 Achilles tendon reflex, 77  
     in tabes dorsalis, 837  
 Achondroplasia, 955  
     of adults, 959  
     in art, 960  
     bones in, 956  
     of children, 959  
     diagnosis of, 967  
     from infantilism, 967  
     epiphyseal cartilages in, 958  
     etiology of, 961  
     gigantism and, 961  
     hands in, 962  
     heredity and, 961  
     macrocephaly in, 963  
     mentality in, 963  
     nature of, 965  
     pathology of, 955  
     periosteal dysplasia, 969  
     prognosis of, 968  
     ribs in, 957  
     rickets and, 966  
     skull in, 956  
     of stillborn, 955  
     symptoms of, 962  
     syphilis and, 967  
     thyroid and, 965  
     treatment of, 968  
     varieties of, 958  
     vertebræ in, 957  
 Achromates, 25  
 Achromatopsia, 271  
 Aeromegaly, diagnosis of, from Paget's  
     disease, 977  
 Acroparesthesia, 32  
 Actinomycoma of brain, 321  
 Acts, anomalies in duration of, 66  
     in intensity of, 66  
 Adductor laryngeal paralysis, 521  
 Adenolipomatosis, symmetrical, 818  
 Adiadokokinesis, 72  
     in brain tumor, 339  
 Adiposis cerebri, 818  
     dolorosa, 817  
     ductless glands in, 818  
     etiology of, 817  
     pain in, 819, 820

Adiposis dolorosa, pathology of, 818  
     prognosis of, 821  
     symptoms of, 819  
     treatment of, 821  
     tuberosa simplex, 818  
 Affective states, anomalies of, 59  
 Agenesis of cerebellum, 287  
 Agitation, affective, 65  
     hallucinatory, 65  
     motor, 63  
 Agnosia, 269  
 Agraphia, 75  
     in aphasia, 297, 298  
     in migraine, 715  
 Akinesia algera, 816  
 Akinetic motor phenomena in hysteria, 678  
 Akro-elephantiasis ossea, 982  
 Albuminuria in infantile convulsions, 795.  
     in polyneuritis, 590  
 Albuminuric retinitis, 475  
 Alcoholic neuritis, 579  
     diagnosis of, from tabes dorsalis,  
         845  
     pseudo-paresis, diagnosis of, from  
         paresis, 860  
 Alcoholism, diagnosis of, from apoplexy,  
     442  
     encephalitis and, 490  
     epilepsy and, 593, 596  
     syphilitic myelitis and, 238  
 Alexia, 54  
     in brain tumor, 333  
 Allochiria, 268  
     in tabes dorsalis, 838  
 Allopsyche, 42  
 Alveolar sarcoma of brain, 314  
 Amaurosis, 34, 480  
 Amaurotic family idiocy, 863  
 Amblyopia, 34, 480  
 Amimia in aphasia, 296  
 Amnesia, 53, 270  
     anterograde, 53  
     circumscribed, 53  
     retarded, 53  
     retrograde, 53  
     retrospective, 53  
 Amnesic aphasia, 54, 74, 298  
 Amyotonia congenita, 893  
     diagnosis of, 894  
     electrical reactions in, 894  
     etiology of, 893  
     muscles in, 894  
     pathology of, 893  
     symptoms of, 894  
     treatment of, 894

- Amyotrophic lateral sclerosis, 89. *See* Sclerosis, amyotrophic lateral.
- syringomyelia, 220
- Anacrusis, 33
- Analgesia, 30
- dolorosa, 30
- in hysteria, 682
- in syringomyelia, 223
- Anarthria, 74
- Anemia of brain, 392
- in hysteria, 688
- in multiple neuritis, 585
- neuralgia and, 726
- retinitis in, 475
- of spinal cord, 255
- subacute combined sclerosis and, 128
- Anesthesia, 28
- in brain tumor, 337
- in compression of spinal cord, 191
- dissociative, in syringomyelia, 223
- in hysteria, 651, 680, 681
- of larynx, 522
- in migraine, 713, 714
- in neuralgia, 728
- in occupation neuroses, 747
- in tabes dorsalis, 838
- tactile, in syringomyelia, 223
- in traumatic neuroses, 651
- Aneurism of cerebral arteries, 398
- diagnosis of, from brain tumor, 344
- Anger, 61
- Angioma of brain, 317
- Annular rickets, 955
- Anomalies of conduct, 63
- in duration of ideas, 52
- in intensity of ideas, 52
- of reflexes, 76
- of sense of pain, 29
- of will, 63
- Anomia, 76
- Anopsia, 34
- Anosmia, 33, 471
- Anterior crural nerve, paralysis of, 557
- Anuria in hysteria, 687
- Apathy, pathological, 61
- Aphasia, 292
- in abscess of brain, 381
- amnesic, 54, 298
- in apoplexy, 457
- of apperception, 292
- in arteriosclerosis of brain, 397
- in brain tumor, 331
- Broca's, 295
- classification of, 294
- compound, 293
- diagnosis of, 303
- extrinsic, 301
- intrinsic, 301
- jargon, 297
- Marie's theories on, 300
- mental defect of, 302
- motor, 75, 292, 295
- cortical, 295
- subcortical, 295
- symptoms of, 296
- optic, 54, 299
- Aphasia in pachymeningitis interna, 165
- sensory, 75, 292, 296
- agraphia in, 297
- apraxia and, 299
- logorrhoea in, 297
- mind blindness in, 299
- subcortical, 299
- symptoms of, 299
- tone deafness in, 297
- Wernicke's, 299
- word blindness in, 298
- deafness in, 297
- subcortical, 75
- summary of, 299
- total, 75
- true, 292
- visual, 298
- Aphemia, 75, 300
- Aplasia, bone, myeloplastic, 978
- Apoplexy, 405
- age and, 407
- aphasia in, 457
- arteriosclerosis and, 396, 427
- aura in, 433
- duration of, 435
- bleeding in, 453
- blood-pressure in, 435
- brain injuries in, 438
- tumor and, 419
- cerebral aneurism and, 399
- diplegia and, 419
- hemorrhages and, 410, 411, 414, 415
- palsies, congenital and, 410
- softening in, 431
- Cheyne-Stokes' breathing in, 436
- childbirth and, 423
- chronic nephritis and, 427
- consciousness in, 432, 433
- in decade I, 409
- II, 421
- III, 423
- IV, 425
- V, 426
- VI, 427
- VII, 428
- VIII, 429
- IX, 429
- diabetes mellitus and, 427
- diagnosis of, 441
- coma in, 450
- convulsions in, 450
- from alcoholic coma, 442
- from brain tumor, 343
- from diabetic coma, 442
- from encephalitis, 375
- from epilepsy, 443
- from hysteria, 443, 446
- from malaria, 444
- from opium poisoning, 442
- from sunstroke, 444
- from trauma, 444
- from uremia, 441
- occupation and, 449
- of vascular lesion, 447
- diphtheria and, 417
- embolism and, 418, 421, 424, 425, 427



- Apoplexy, embolism and, diagnosis of, 448  
 epilepsy and, 421  
 etiology of, 406  
 exciting causes of, 430  
 eyes in, 439  
 eye-grounds in, 437  
 fever in, 437  
 hemiplegia and, 412, 457  
     congenital and, 409  
     etiology of, 413  
 hemorrhage and, 422, 424, 427, 428, 431  
     diagnosis of, 448, 450  
 hysteria and, 419  
 Little's disease and, 419  
 measles and, 417  
 occurrence of, 406  
 onset of, 431  
 paralysis in, 438, 440  
 paralytic conjugate deviation in, 439  
 porencephaly and, 409  
 pregnancy and, 423, 425  
 prognosis of, 451  
 pulse in, 835  
 pupils in, 439  
 race and, 407  
 reflexes in, 439  
 respiration in, 436  
 rupture of aneurism and, 429  
 scarlet fever and, 417  
 sequels of, 456  
 sex and, 407  
 spastic conjugate deviation in, 438  
 symptoms of, 429  
 syphilis and, 423, 425, 426, 427  
     hereditary and, 418  
 thrombosis and, 423, 425  
     diagnosis of, 448  
 toxemia and, 425  
 trauma and, 424  
 treatment of, 452  
     prophylactic, 452  
     surgical, 454  
 typhoid fever and, 417, 425  
 whooping cough and, 417
- Apraxia, 66, 75, 269  
 aphasia and, 299  
     in diseases of corpus callosum, 276
- Arachnoidal cysts of brain, 318
- Argyll-Robertson pupil, 487  
     in general paresis, 856  
     in tabes dorsalis, 836
- Arrhythmic movements in hysteria, 677
- Arm, lower, paralysis of, 552  
     upper, paralysis of, 551
- Arteries, carotid, anatomy of, 388  
     cerebral, aneurism of, 398  
         embolism of, 400  
         obliteration of, 400  
         rupture of, 400  
         thrombosis of, 400  
     vertebral, anatomy of, 387
- Arteriosclerosis, apoplexy and, 427  
     of brain, 393  
     cerebral aneurism and, 399  
         thrombosis and, 400  
     neuralgia and, 727
- Arteriosclerosis in Paget's disease, 973
- Arteriosclerotic dementia, diagnosis of, from paresis, 861
- Artery, basilar, anatomy of, 387
- Arthralgia in hysteria, 683
- Arthritis deformans, 895  
     age and, 899  
     atrophic, 920  
         bones in, 907  
         diagnosis of, 932  
         prognosis of, 934  
         symptoms of, 920  
         treatment of, 942  
     bones in, 904, 906  
     cartilage in, 904, 906  
     classification of, 898  
     deformity in, prevention of, 941  
         treatment of, 942  
     diagnosis of, 930  
         from gonorrhœal arthritis, 931  
         from gout, 931  
         from osteo-arthritis, 987  
         from rheumatic fever, 930  
     diet in, 938  
     effusion in, 904  
     endocarditis in, 914  
     etiology of, 899  
     gout and, 930  
     heredity and, 901  
     hygiene in, 938  
     hypertrophic, 920  
         bones in, 907  
         diagnosis of, 932  
         Heberden's nodes in, 921  
             treatment of, 943  
     hip-joints in, 922  
     knee-joint in, 923  
     metacarpophalangeal joints in, 922  
     pain in, 921, 922  
     prognosis of, 935  
     sacro-iliac joint in, 923  
     symptoms of, 920  
     treatment of, 942  
     infections and, 900, 902  
     influenza and, 901  
     intestinal infection and, 903  
     joints in, treatment of, 939  
     massage in, 938, 940  
     menopause and, 902  
     metabolism and, 903, 907  
     monarticular, 919, 934  
         diagnosis of, 932  
     muscles in, 907  
     nature of, 898  
     nervous system in, 903, 907  
     pathology of, 904  
     peri-articular, 908  
         acute, 909  
         in advanced life, 918  
         arthritis in, 910  
         atrophy in, 914  
         blood in, 916  
         character of attack in, 908  
         in children, 917  
         chronic, 909

- Arthritis deformans, peri-articular, circulatory system in, 914  
     counterirritation in, 941  
     distribution of lesions in, 913  
     exercises in, 940  
     fever in, 909, 914  
     glandular enlargement in, 909, 915  
     hip-joint in, 913  
     leukocytosis in, 916  
     œdema in, 917  
     onset of, 908  
     pain in, 909, 910  
     pericarditis in, 914  
     pleurisy in, 915  
     prognosis of, 933  
     pulse in, 909, 914  
     reflexes in, 917  
     respiratory tract in, 915  
     shoulder-joint in, 911  
     skin in, 916  
     spleen in, 909, 915  
     subcutaneous nodules in, 915  
     symmetry in, 913  
     symptoms of, 908  
     treatment of, 940  
     urine in, 916  
     pregnancy and, 902  
     prognosis of, 933  
     pyorrhœa alveolaris and, 901  
     race and, 900  
     Raynaud's disease and, 929  
     scleroderma and, 929  
     sex and, 899  
     of spinal joints, 910  
     spondylitis in, 923  
     Still's disease and, 917, 931  
     symptoms of, 908  
     synovial membrane in, 904, 905  
     tonsillitis and, 901  
     trauma and, 901  
     treatment of, 936  
     vaccines in, 939  
     villous arthritis in, 929  
   in hypertrophic osteo-arthritis, 985  
   infectious, 896  
   rheumatoid, 897, 908  
   villous, 929  
     treatment of, 942  
 Arthropathy in syringomyelia, 225  
   in tabes dorsalis, 842  
 Ascending multiple neuritis, 576  
 Association, anomalies of, 46  
   tests, 48  
 Astasia-abasia, 815  
   affective, 816  
   diagnosis of, 816  
     from akinesia algesia, 816  
     from myasthenia gravis, 816  
   emotional, 816  
   etiology of, 815  
   fatigue in, 816  
   hypochondriacal, 816  
   hysteria and, 680, 816  
   prognosis of, 817  
   psychasthenic, 816  
 Astasia-abasia, symptoms of, 815  
   treatment of, 817  
 Astereognosis, 30, 268  
   in lesion of optic thalamus, 281  
   in tabes dorsalis, 838  
 Asthenic paralysis, 798  
 Asthenopia, 36  
 Astigmatism, 36  
 Asyllabia, 298  
 Asymbolia, 269  
   optic, 54  
 Asynergy in brain abscess, 381  
   tumor, 339  
   in lesion of cerebellum, 286  
 Ataxia, 71  
   in abscess of brain, 381, 384  
   in brain tumor, 336, 339  
   cerebellar, 72  
     Marie's hereditary, 130, 136  
   in disseminated sclerosis, 147, 154  
   Friedreich's, 130  
     ataxia in, 135  
     diagnosis of, 138  
       from disseminated sclerosis, 138  
       from Huntington's chorea, 139  
       from tabes dorsalis, 138  
   etiology of, 131  
   gait in, 136  
   nystagmus in, 135  
   pain in, 136  
   pathology of, 131  
   prognosis of, 139  
   symptoms of, 133  
   treatment of, 139  
   vasomotor paresis in, 136  
   in hysteria, 652  
   in lesion of cerebellum, 287  
     of mid-brain, 284  
     of optic thalamus, 280  
   in multiple neuritis, 583  
   in tabes dorsalis, 839  
 Athetoid movements, 70  
 Athetosis in cerebral diplegia, 420  
   in lesion of optic thalamus, 280  
 Athymia, 61  
 Atony of muscles, 68  
 Atrophic arthritis deformans, 920  
 Atrophy of cerebellum, 287  
   facial, in migraine, 719  
   muscular, 68  
   in arthritis deformans, 914  
   in diseases of peripheral nerves, 534  
   progressive, Charcot-Marie-Tooth type, 109  
     diagnosis of, from syringomyelia, 226  
     neural, 109  
       contractures in, 110  
       diagnosis of, 111  
       pathology of, 109  
       reflexes in, 110  
       symptoms of, 109  
   spinal, 80  
   age and, 81

Atrophy, muscular, progressive spinal,  
     atrophy in, 84  
     of childhood, 87  
     atrophy in, 88  
     course of, 88  
     diagnosis of, 88  
     familial, 87  
     hereditary, 87  
     paresis in, 88  
     pathology of, 88  
     symptoms of, 88  
     tremors in, 88  
     definition of, 80  
     diagnosis of, 86  
         from progressive  
         neural muscular,  
         111  
     electrical reactions in,  
         85  
     etiology of, 81  
     heredity and, 81  
     paralysis in, 84  
     pathology of, 82  
     pregnancy and, 81  
     prognosis of, 86  
     reflexes in, 85  
     symptoms of, 83  
     treatment of, 87  
     tremors in, 85  
     weakness in, 84  
     type, Duchenne-Aran, 80  
         Werdnig-Hoffmann, 87  
     in occupation neuroses, 744  
     olivopontocerebellar, 287  
     optic, 37, 478  
         in tabes dorsalis, 841  
     in osteo-arthropathy, 986  
     in progressive muscular dystrophy,  
         119  
     of tongue, 531  
 Attention, anomalies of, 45  
 Attonnity; 65  
 Auditory nerve, diseases of, 506  
     vertigo, 512  
 Aura in epilepsy, 600  
 Aural vertigo, 513  
 Automatism, 44  
 Autopsyche, 43

## B

BABINSKI's hypothesis of hysteria, 658  
     phenomenon, 78  
     sign in apoplexy, 440  
         in disseminated sclerosis, 149  
 Bacteria, multiple neuritis and, 575, 576  
     myelitis and, 229  
 Bacteriological examination of cerebro-  
     spinal fluid, 177  
 Bacteriology of chorea, 778  
     of encephalitis, 362  
     of infective myelitis, 231  
     of Landry's paralysis, 248  
 Basal ganglia, tumors of, 334  
 Basilar artery, anatomy of, 387  
 Bathyanesthesia, 30  
 Bathyesesthesia, 17

Benedikt's syndrome, 369  
 Biological hypothesis of hysteria, 665  
 Birth palsies, brachial, 552  
     trauma, epilepsy and, 594  
 Bladder in hemiplegia, 467  
     in syphilitic myelitis, 240  
     in tabes dorsalis, 840  
 Bleeding in apoplexy, 453  
     in hysteria, 687  
 Blind spot, 34  
 Blindness, 34  
     in amaurotic family idiocy, 869  
     cortical, 54  
     day, 480  
     in hysteria, 684  
     mind, 53, 271  
     night, 480  
     psychic, 299  
     word, 54  
 Blood in arthritis deformans, 916  
     in hysteria, 688  
     in osteomalacia, 952  
 Bloodvessels of brain, 387  
     in syphilitic myelitis, 238  
 Bones in achondroplasia, 956  
     in arthritis deformans, 904, 906, 907  
     in facial hemiatrophy, 992  
     in osteogenesis imperfecta, 979  
     in osteomalacia, 949  
     in Paget's disease, 971  
     in progressive myositis ossificans, 883  
     in syringomyelia, 225  
 Brachial neuralgia, 737  
     neuritis, 553  
     plexus, paralysis of, 550  
         prognosis of, 553  
 Bradyllalia, 74  
 Brain, abscess of, 378  
     aphasia in, 381  
     asynergy in, 381  
     ataxia in, 381, 384  
     bronchiectasis and, 379  
     convulsions in, 380, 381  
     diagnosis of, from acute menin-  
         gitis, 172, 386  
         from sinus thrombosis, 386  
         from tumor, 344, 386  
     empyema and, 379  
     etiology of, 378  
     eye-grounds in, 380  
     fever in, 380  
     headache in, 379  
     hemianopsia in, 381, 382, 384  
     hemiataxia in, 381, 384  
     hemiparesis in, 384  
     idiopathic, 385  
     infected wounds and, 379  
     metastatic, 384  
     nystagmus in, 384  
     otitic, 383  
     pain in, 381, 384  
     paralysis in, 381  
     paraphasia in, 381  
     pathology of, 378  
     prognosis of, 386  
     pulse in, 380  
     respiration in, 380

- Brain, abscess of, rhinogenic, 385  
 sensation in, 381  
 sinus thrombosis and, 379  
 symptoms of, 379  
   cerebellar, 381  
   focal, 380  
     motor, 381  
   frontal, 381  
   occipital, 382  
   temporal, 381, 383  
 temporal lobe, 383  
 traumatic, 382  
   acute, 382  
   diagnosis of, 382  
   latent, 382  
 treatment of, 386  
 vertigo in, 379, 384  
 vomiting in, 379  
 word blindness in, 382  
 actinomycoma of, 321  
 acute red softening of, 359  
 in amaurotic family idiocy, 864  
 anemia of, 392  
   Cheyne-Stokes respiration in, 392  
   convulsions in, 392  
   headache in, 393  
   symptoms of, 392  
 angioma of, 317  
 arteries of, anastomosis of, 387  
   aneurism of, 398  
     apoplexy and, 399  
     arteriosclerosis and, 399  
     choked disk in, 399  
     dizziness in, 399  
     headache in, 399  
     syphilis and, 399  
     vomiting in, 399  
 embolism of, 400  
   age and, 401  
   etiology of, 401  
   pathology of, 403  
 obliteration of, 400  
 rupture of, 400  
 thrombosis of, 400  
   age and, 400  
   arteriosclerosis and, 400  
   chlorosis and, 400  
   etiology of, 400  
   pathology of, 403  
   syphilis and, 400, 401  
   typhoid fever and, 400  
 arteriosclerosis of, 393  
   age and, 394  
   aphasia in, 397  
   apoplexy in, 396  
   convulsions in, 397  
   diagnosis of, 398  
     from paresis, 398  
   etiology of, 393  
   headache in, 396  
   neurasthenia in, 395  
   pathology of, 394  
   psychoses in, 397  
   symptoms of, 394  
     mental, 397  
   syphilis and, 393  
   treatment of, 398
- Brain, arteriosclerosis of, vertigo in, 396  
 bloodvessels of, 387  
   anatomy of, 387  
   diseases of, 387  
   physiology of, 389  
 carcinoma of, 315  
 centre of hearing in, 271  
   of smell in, 271  
 centrum semiovale, lesion of, 275  
 cerebellum, lesion of, 284  
 chloroma of, 315  
 cholesteatoma of, 316  
 chordoma of, 317  
 convolutions of, central, lesions of, 274  
   paralysis in, 274  
   spasm in, 275  
 corpora striata, lesion of, 277  
 corpus callosum, lesion of, 276  
 cortex of, motor area of, 265, 273  
   diseases of, focal symp-  
     toms of, 266  
   sensory area of, 265, 273  
   diseases of, focal symp-  
     toms of, 267  
     localization of, 273  
 crura cerebri, lesion of, 281  
 cysticercus cellulosus of, 318  
 cystoglioma of, 313  
 cystosarcoma of, 315  
 cysts of, 318  
   arachnoidal, 318  
   congenital, 318  
   echinococcus, 319  
   epilepsy and, 599  
   hydatid, 319  
   parasitic, 318  
   simple, 318  
   traumatic, diagnosis of, from  
     tumor, 346  
 disease of, diagnosis of, from hysteria,  
   692  
   topical diagnosis of, 265  
 enchondroma of, 317  
 endothelioma of, 313  
   diagnosis of, from neurofibroma,  
     314  
   dural, 313  
 epithelioma of, 315  
 fibroma of, 317  
   acoustic, 314  
 glioma of, 311  
   diagnosis of, 312  
   diffuse, 312  
   ependymal, 311  
 granuloma of, infectious, 319  
 gumma of, 320  
 hemorrhage in, 401  
 hyperemia of, 392  
 inflammation of, 359  
 injuries of, in apoplexy, 438  
 internal capsule, lesion of, 276  
 lenticular nucleus, lesion of, 278  
   zone, lesion of, 278  
 lipoma of, 317  
 myeloma of, 315  
 myxosarcoma of, 315  
 neuroblastoma of, 317

- Brain, neurocystoma of, 317  
 neurofibroma of, diagnosis of, from  
 endothelioma, 314  
 neuroglioma of, 311  
 neuroma of, 317  
 optic thalamus, lesion of, 279  
 osteosarcoma of, 314  
 papilloma of, 315  
 pons, lesion of, 289  
 pontomedullocerebellar space, lesion  
 of, 289  
 psammoma of, 318  
 psammosarcoma of, 315  
 quadrigeminal bodies, lesion of, 282  
 sarcoma of, 313, 314  
   alveolar, 314  
   diffuse, 315  
   perforating, 314  
 sclerosis of, 140  
 stem, tumors of, 335  
 syphilis of, 823. *See* Syphilis, cere-  
 brospinal.  
 syphiloma of, 320, 321  
 tegmentum, lesion of, 281  
 teratoma of, 316  
 tubercle of, solitary, 319  
 tuberculoma of, 319  
 tuberculous meningo-encephalitis of,  
 319  
 tumor of, 308  
   adiadokokinesis in, 339  
   age and, 309  
   alexia in, 333  
   anesthesia in, 337  
   aphasia in, 331  
   asynergia in, 339  
   ataxia in, 336, 339  
   in basal ganglia, 334  
   in brain stem, 335  
   cachexia hypophyseopriva in, 341  
   in cavum Meckelii, 340  
   in centrum semiovale, 334  
   in cerebellum, 338  
   cerebrospinal fluid in, 323  
   choked disk in, 326, 338  
     labyrinth in, 328  
   convulsions in, 329, 333  
   in corpora quadrigemina, 336  
   in corpus callosum, 335  
   course of, 346  
   in cranial base, 340  
   in crura, 335  
   decompression in, 349  
   dermoid, 316  
   diagnosis of, 330, 342  
     from abscess, 344, 386  
     from aneurism, 344  
     from apoplexy, 343  
     from blastomycotic menin-  
     gitis, 345  
     from cerebral thrombosis,  
     343  
     from disseminated sclerosis,  
     156, 343  
     from encephalitis, 374  
     from ependymitis, 345  
     from general paresis, 343  
     from hydrocephalus, 345  
     from hysteria, 342  
     from lead encephalopathy,  
     346  
     from nephritis, 344  
     from pseudo-tumor, 346  
     from serous meningitis, 345  
     from tabes dorsalis, 345  
     from traumatic cysts, 346  
     from vascular lesions, 343  
     lumbar puncture in, 330  
     regional, 330  
   dislocation from pressure in, 323  
   dizziness in, 329, 338  
   drowsiness in, 329  
   dyschromatopsia in, 329  
   dysesthesia in, 339  
   epilepsy in, 331, 333, 599  
   etiology of, 309  
   in fourth ventricle, 338  
   in frontal lobe, 331  
   glycosuria in, 338, 342  
   headache in, 324, 338  
   hemiachromatopsia in, 334  
   hemianopsia in, 332, 334  
   hemiplegia and, 419  
   hydrocephalus in, 338  
   in hypophysis cerebri, 341  
   hypopituitarism in, 341  
   incidence of, 308  
   in medulla oblongata, 337  
   Ménière's syndrome in, 337  
   migraine and, 720  
   nystagmus in, 336, 339, 340  
   in occipital lobe, 334  
   oedema in, 323  
   pain in, 338  
   in paracentral convolutions, 332  
   paralysis in, 337  
   in parietal lobe, 333  
   pathological physiology of, 321  
   pathology of, 310  
   in pineal gland, 336  
   polydipsia in, 338  
   polyuria in, 337, 338, 342  
   in pons, 336  
   prognosis of, 346  
   psychic disturbances in, 329,  
   342  
   salivation in, 332, 337  
   sex and, 309  
   symptoms of, 321  
     focal, 329  
     general, 322  
   in tegmentum, 336  
   in temporal lobe, 331  
   thalamic syndrome in, 334  
   trauma and, 309  
   treatment of, 347  
   tremors in, 340  
   vertigo in, 329, 338  
   vomiting in, 326, 338  
 Broca's aphasia, 75, 295  
 Bromides in epilepsy, 620  
 Bronchiectasis, abscess of brain and, 379

Bronchiectasis, hypertrophic osteo-arthropathy and, 982  
 Brown-Séquard paralysis, 185, 204  
 Bulbar changes in amyotrophic lateral sclerosis, 92, 100  
   muscles in myasthenia gravis, 802  
   myelitis, acute, 368  
   palsy, acute inflammatory, 368  
     progressive, 113  
   paralysis, ascending, 798.

## C

CACHEXIA hypophyseopriva in brain tumor, 341  
   multiple neuritis and, 576  
 Calcium, tetany and, 751  
 Capsule, internal, lesion of, 276  
   conjugate deviation of eyes in, 277  
   hemianesthesia in, 276  
   hemianopsia in, 276  
   hemicontracture in, 277  
   hemiplegia in, 276  
   hemithetosis in, 277  
   paralysis in, 276  
 Carcinoma of brain, 315  
   of spinal cord, 196  
 Carcinomatosis, diagnosis of, from cerebrospinal syphilis, 827  
 Cardiac crises in tabes dorsalis, 842  
   epilepsy, 597  
   nerves, paralysis of, 522  
 Cardialgia in hysteria, 683  
 Caries, sicca, 189  
   spinal, 188. *See* Spinal caries.  
 Carotid arteries, anatomy of, 388  
 Cartilage in arthritis deformans, 904, 906  
 Catatonic rigidity, 65  
 Cauda equina, diseases of, diagnosis of, 186  
   from paralysis of lower limbs, 561  
   from sciatica, 565  
 Causalgia in diseases of peripheral nerves, 539  
 Cavum Meckelii, tumors of, 340  
 Centrum semiovale, lesion of, 275  
   epileptoid attacks in, 275  
   speech in, 275  
   vision in, 275  
   tumor in, 276, 334  
 Cephalocele, hydrocephalus and, 354  
 Cephalomegalia, 988  
 Cerebellar artery, posterior inferior, occlusion of, 288  
   ataxia in, 288  
   paralysis in, 288  
   sympathetic in, 288  
   vertigo in, 288  
   ataxia, 72  
     gait in, 74  
     Marie's hereditary, 130, 136  
   deficit, syndrome of, 287  
   localization, 286  
   symptoms in migraine, 716  
   syndrome, 286

Cerebellum, agenesis of, 287  
   atrophy of, 287  
   functions of, 284  
   lesion of, 284  
     asynergia in, 286  
     ataxia in, 287  
     gait in, 286  
     hypotonia in, 287  
     paralysis in, 286  
     reflexes in, 286  
     symptoms of, 285  
   sclerosis of, 287  
   tumors of, 338  
 Cerebral arteries, aneurism of, 398  
   embolism of, 400  
   obliteration of, 400  
   rupture of, 400  
   thrombosis of, 400  
 arteriosclerosis, 393. *See* Brain, arteriosclerosis of.  
 bloodvessels, diseases of, 387  
 diplegia, 419  
 disseminated sclerosis, 154  
 hemiplegia, congenital, 409  
   infantile, 412  
 hemisphere, ganglia of, 278  
 hemorrhage, 401, 410, 411, 414, 415  
   age and, 403  
   diagnosis of, from encephalitis, 375  
   etiology of, 401  
   pathology of, 405  
 localization hypothesis of hysteria, 655  
 nerves, diseases of, 471  
 palsies, congenital, 410  
 softening in apoplexy, 431  
   pathology of, 403  
 thrombosis, diagnosis of, from brain tumor, 343  
 toxemia, diagnosis of, from encephalitis, 375  
 Cerebrospinal disseminated sclerosis, 154  
   fever, hydrocephalus and, 357  
   fluid, 351  
     circulation of, 352  
     examination of, 174  
       bacteriological, 177  
       chemical, 176  
       cytological, 176  
     lymphocytosis of, 177  
     pressure of, 175  
     in syphilis, 828  
   meningitis, acute, 167  
   syphilis, 823  
     diagnosis of, from disseminated sclerosis, 156  
   tuberculosis, diagnosis of, from syphilis, 827  
 Cervical plexus, diseases of, 542  
   paralysis of, 543  
   ribs, 554  
     cyanosis in, 556  
     diagnosis of, 556  
     gangrene in, 556  
     pain in, 555  
     pulse in, 556

- Cervical ribs, symptoms of, 555  
 thrombosis in, 556  
 treatment of, 556
- Cervico-occipital neuralgia, 736
- Charcot-Marie-Tooth type of progressive muscular atrophy, 109
- Cheirokinesthetic agraphia, 75
- Cheyne-Stokes respiration in acute meningitis, 170  
 in apoplexy, 436
- Chloroma of brain, 315
- Chlorosis, cerebral thrombosis and, 400
- Choked disk, 36  
 in brain tumor, 326, 338  
 in cerebral aneurism, 399  
 mechanism of, 476  
 stages of, 327
- Cholesteatoma of brain, 316
- Chondrodystrophia fetalis hypoplastica, 958  
 malacica, 959
- Chordoma of brain, 317
- Chorea, acute, 774  
 age and, 774  
 bacteriology of, 778  
 climate and, 774  
 complications of, 781  
 course of, 781  
 definition of, 774  
 diagnosis of, from hysteria, 693  
 emotions and, 774  
 endocarditis and, 776, 781  
 etiology of, 774  
 reflex, 777  
 fever in, 779  
 heredity and, 775  
 herpes zoster in, 781  
 infections and, 777  
 malignant, 779  
 mild, 778  
 paralytic phenomena in, 780  
 pathogenesis of, 775  
 pathology of, 778  
 pregnancy and, 777, 779  
 prognosis of, 781  
 psychic manifestations in, 780  
 race and, 774  
 reflexes in, 781  
 rheumatic fever and, 775  
 severe, 779  
 sex and, 774  
 symptoms of, 778  
 synonyms of, 774  
 syphilis and, 775, 777  
 treatment of, 782  
 urine in, 781
- chronic hereditary, 787  
 degenerative, 787  
 electric, 787  
 habit, 784  
 definition of, 784  
 diagnosis of, 786  
 etiology of, 785  
 symptoms of, 785  
 treatment of, 786
- Huntington's, 787  
 diagnosis of, 789
- Chorea, Huntington's, etiology of, 787  
 heredity and, 788  
 pathology of, 788  
 prognosis of, 790  
 symptoms of, 788  
 psychic, 789  
 treatment of, 790
- insaniens, 779  
 minor, 774  
 mollis, 780  
 posthemiplegic, 466  
 progressive, 787  
 Sydenham's, 774
- Choreiform movements, 70
- Choroidoretinitis, syphilitic, 475
- Chvostek's phenomenon, 71  
 sign in tetany, 758
- Circulatory disturbances, neurasthenia and, 631  
 system in arthritis deformans, 914
- Circumflex nerve, paralysis of, 546
- Circumspection, 43
- Clubbed fingers, 981
- Coccygeal plexus, neuralgia of, 741
- Cochlear nerve, diseases of, 506  
 symptoms of, 507  
 treatment of, 508
- Color blindness, 25
- Coma, diagnosis of, 441  
 in lesion of pons, 290
- Compression myelitis, 229
- Confusion, 43
- Congenital cerebral hemiplegia, 409  
 cysts of brain, 318  
 heart disease, convulsions and, 794  
 osteosclerosis, 968  
 rickets, 955
- Conjugate deviation in apoplexy, 438, 439
- Consciousness, allopsyche and, 42  
 anomalies of, 41  
 autopsyche and, 43  
 somatopsyche and, 42
- Contractures in amyotrophic lateral sclerosis, 99  
 in facial paralysis, 502  
 in hemiplegia, 464  
 in hereditary spastic spinal paralysis, 106  
 in hysteria, 651  
 of muscles, 68  
 in progressive neural muscular atrophy, 110  
 in traumatic neuroses, 651
- Convulsions, central, lesions of, 274
- Convulsions in abscess of brain, 380, 381  
 in acute meningitis, 170  
 in anemia of brain, 392  
 in arteriosclerosis of brain, 397  
 in brain tumor, 329, 333  
 in general paresis, 857  
 hemiplegia and, 415  
 in hysteria, 673  
 infantile, 790  
 age and, 790  
 albuminuria in, 795  
 alcohol and, 793  
 congenital heart disease and, 794

- Convulsions, infantile, consciousness in,  
     loss of, 795  
     diagnosis of, 795  
         from epilepsy, 796  
     epilepsy and, 594  
     etiology of, 790  
     infections and, 793  
     motor phenomena in, 794  
     onset of, 794  
     opisthotonos in, 794  
     paralysis in, 795  
     predisposition to, 791  
     prognosis of, 796  
     reflex irritation and, 792  
     respiration in, 794  
     rickets and, 792, 793  
     symptoms of, 794  
     thymus and, 794  
     treatment of, 796  
     whooping cough and, 793  
     in lesion of pons, 290  
     in pachymeningitis interna, 165  
     postnatal infantile hemiplegia and,  
         415  
     Convulsive tic, 784  
     Coordination, disturbances of, 71  
     Corpora quadrigemina, tumors of, 336  
         striata, lesion of, 278  
     Corpus callosum, disease of, 276  
         tumor of, 276, 335  
     Cortex of brain, motor area of, 265, 273  
         sensory area of, 265, 273  
     Cortical blindness, 54  
         deafness, 33  
     Coryza in migraine, 718  
     Cramps in occupation neuroses, 744  
         professional, 741  
     Cranial base, tumors of, 340  
         nerves in tabes dorsalis, 841  
     Cremaster reflex, anomalies of, 78  
     Crura cerebri, lesion of, 281  
         tumors of, 335  
     Crural neuralgia, 740  
     Cutaneous reflexes, anomalies of, 77  
     Cyanosis in cervical ribs, 556  
         hypertrophic osteo-arthritis and,  
             982  
     Cystoglioma of brain, 313  
     Cystosarcoma of brain, 315  
     Cysts of brain, 318  
     Cytological examination of cerebrospinal  
         fluid, 176

## D

- DAY blindness, 480  
     Deafness in cerebrospinal syphilis, 825  
         mind, 54  
         word, 54  
     Deep reflexes, anomalies of, 77  
     Deformity in compression of spinal cord,  
         190  
     Degeneration, progressive lenticular, 278  
         reaction of, 72  
     Degenerative myelitis, 236  
     Delirium in acute meningitis, 170  
         acutum, 66  
     Delusions, 56  
         anomalies of conduct due to, 67  
         depressive, 58  
         distinction from defects in judgment,  
             58  
         expansive, 58  
         in general paresis, 855  
         of grandeur, 57, 855  
         of persecution, 58  
         primary, 56  
         secondary, 56  
     Dementia, anomalies of conduct and, 67  
         præcox, hysteria and, 694, 695  
     Depression, pathological, 60  
     Dercum's disease, 817  
     Dermatitis, bullous, in hysteria, 686  
         in dermatomyositis, 877  
     Dermatomyositis, 875  
         dermatitis in, 877  
         diagnosis of, 878  
         distribution of, 875  
         etiology of, 875  
         fever in, 877  
         nervous system in, 877  
         cedema in, 877  
         pain in, 877  
         pathology of, 876  
         prognosis of, 878  
         symptoms of, 876  
         treatment of, 878  
     Dermoid tumors of brain, 316  
     Development, mental, trauma and, 650  
     Diabetes, coma of, diagnosis of, from  
         apoplexy, 442  
         mellitus, apoplexy and, 427  
     Diadokokinesis, 72  
     Diaphragmatic neuralgia, 737  
     Diarrhœa in hysteria, 688  
         in migraine, 718  
     Dichromates, 25  
     Diffuse sclerosis, 160  
     Diphtheria, apoplexy and, 417  
         multiple neuritis and, 575  
     Diplegia, cerebral, 419  
         athetosis in, 420  
         epilepsy and, 421  
         symptoms of, 419  
         congenital, diagnosis of, from amaurotic family idiocy, 869  
     Diplopia in disseminated sclerosis, 150  
         monocular, 480  
         in myasthenia gravis, 802  
     Dislocation from pressure in brain tumors,  
         323  
     Disorientation, 44  
     Disseminated myelitis, 235  
         sclerosis, 140  
     Dissociation, 51  
     Dizziness in brain tumor, 329, 338  
     Dorsodynia, 945  
     Dream state, 689  
     Drowsiness in brain tumor, 329  
     Duchenne-Erb palsy, 551  
     Dura mater, diseases of, 163  
         hematoma of, diagnosis of, from  
             brain tumor, 345  
     Dysacusis, 33



Dysarthria, 74  
 Dyschiria, 268  
 Dyschondroplasia, 969  
 Dyschromatopsia in brain tumor, 329  
 Dysesthesia, 32  
   in brain tumor, 339  
 Dysphagia in brain tumor, 337  
 Dysphonia, spastic, 522  
 Dysplastic periosteal achondroplasia, 969  
 Dyspnoea in Landry's paralysis, 251  
   in myasthenia gravis, 804  
   in paralysis of cervical plexus, 543  
 Dyspraxia, 75  
 Dysthymia, 60  
 Dystrophy, muscular, bulbar, diagnosis of,  
   from bulbar palsy, 117  
   progressive, 117  
     atrophy in, 119  
     diagnosis of, 123  
       from anterior poliomye-  
       litis, 124  
       from myasthenia gravis,  
       124  
       from progressive neural  
       muscular atrophy, 111  
     etiology of, 117  
     facio-scapulo-humeral, 122  
     hereditary, 123  
     infantile, 123  
     pathology of, 118  
     prognosis of, 124  
     pseudo-hypertrophic, 122  
     reflexes in, 121  
     symptoms of, 119  
     treatment of, 124  
     types of, 122  
     weakness in, 120  
     of Zimmerlin, 123

## E

ECHINOCOCCUS cysts of brain, 319  
 Echolalia, 65  
 Echopraxia, 65  
 Elbow-joint in arthritis deformans, 911  
 Electric chorea, 787  
 Electrical hyperexcitability in tetany, 757  
   reactions in diseases of peripheral  
     nerves, 535  
     in multiple neuritis, 582  
     in progressive spinal muscular  
     atrophy, 85  
 Elephantiasis neuromatosa, 571  
 Embolism, apoplexy and, 418, 422, 424,  
   427, 428, 431  
   of cerebral arteries, 400  
   postnatal infantile hemiplegia and,  
   418  
 Emmetropia, 36  
 Emotional states, anomalies of, 59  
 Emotions, 59  
 Empyema, abscess of brain and, 379  
 Encephalic tuberculoma of brain, 319  
 Encephalitis, 359  
   acute, 360, 361  
     hemorrhagic, 367  
   Encephalitis, acute hyperplastic, 370  
     non-suppurative, 370  
   in advanced years, 375  
   bacteriology of, 362  
   chronic, 361  
   classification of, 360  
   congenita, 361, 370  
   definition of, 359  
   diagnosis of, 373  
     from apoplexy, 375  
     from brain tumor, 374  
     from bulbar palsy, 117  
     from cerebral hemorrhage, 375  
     toxemia, 375  
     from hysteria, 375  
     from sinus thrombosis, 374  
     from syphilis of brain, 375  
   epilepsy and, 599  
   etiology of, 361  
   experimental, 371  
   in infancy and childhood, 376  
   influenzal, 362  
   neonatorum, 370  
   postnatal infantile hemiplegia and,  
   416  
   prognosis of, 377  
   progressive, 361  
   pyogenic, 363  
     age and, 364  
     course of, 364  
     diagnosis of, 364  
     onset of, 364  
     pain in, 363  
     staphylococcus in, 363  
     symptoms of, 364  
   subacute, 360, 361  
   superior, 490  
     alcoholism and, 490  
   synonyms of, 359  
   treatment of, 377  
 Encephalomalacia, pathology of, 403  
 Encephalomyelitis, acute, 369  
   Benedikt's syndrome in, 369  
   diagnosis of, 370  
   Millard-Gubler syndrome in, 369  
   ophthalmoplegia in, 369  
   Weber's syndrome in, 369  
 Encephalopathy, lead, diagnosis of, from  
   brain tumor, 346  
 Enchondroma of brain, 317  
 Endarteritis in cerebrospinal syphilis, 824  
 Endocarditis in arthritis deformans, 914  
   chorea and, 776, 781  
 Endothelioma of brain, 313  
 Enteralgia in hysteria, 683  
 Ependymal glioma of brain, 311  
   inflammation, hydrocephalus and, 358  
 Ependymitis, diagnosis of, from brain  
   tumor, 345  
 Epieritic sensibility, 17  
 Epigastric reflexes, 78  
 Epilepsy, 592  
   affect, 610  
   alcoholism and, 593, 596  
   apoplexy and, 421  
   atypical, 606  
   aura in, 600

- Epilepsy, behavior in, 612  
 birth trauma and, 594  
 in brain tumor, 331, 333  
 bromides in, 620  
 cardiac, 597  
 cerebral diplegia and, 421  
 cysts of brain and, 599  
 definition of, 592  
 degeneration in, 603  
 diagnosis of, 613  
   from apoplexy, 443  
   from hysteria, 614, 691  
   from infantile convulsions, 796  
 diet in, 618  
 disposition in, 611  
 drugs in, 620  
 encephalitis in, 599  
 epileptoid conditions and, 607  
 etiology of, 593  
 exhaustion in, 607  
 experimental seizures in, 609  
 familial, 594  
 hydrocephalus and, 599  
 hydrotherapy in, 619  
 hygiene in, 616  
 hysteroid seizures in, 610  
 idiopathic, 592  
 imbecility and, 594  
 incomplete, 605  
 infantile convulsions and, 594  
 Jacksonian, 592  
 latent, 592  
 meningitis in, 599  
 menstrual, 598  
 migraine and, 615, 711  
 mortality in, 593  
 nocturnal, 598, 610  
 pathology of, 598  
 petit mal, 604  
 porencephaly in, 598  
 psychic, 602  
   alteration in, 602  
   influences and, 597  
 psychical anomalies of, 611  
 psychosis in, 603  
 rationale of seizures in, 608  
 reflex, 596  
 rudimentary, 605  
 seizures in, 601  
 simulation of, 615  
 spasmophilia and, 604  
 speech in, 612  
 stages of, 601  
 status epilepticus in, 609  
 stupor in, 607  
 symptoms of, 600  
 synonyms of, 592  
 syphilis and, 595  
 toxins and, 594  
 trauma and, 596  
 treatment of, 615, 616  
   mental, 619  
   operative, 623  
 true, 592  
 tumors of brain and, 599  
 vasomotor disturbances in, 606  
 will in, 612
- Epileptic attacks in disseminated sclerosis, 153  
   in pseudo-sclerosis, 159  
   insanity, traumatism and, 647  
   psyche, 598, 612
- Epileptoid attacks in lesion of centrum semiovale, 275
- Epiphyseal cartilages in achondroplasia, 958
- Epistaxis in polymyositis hemorrhagica, 879
- Epithelioma of brain, 315
- Equilibrium, sense of, 18
- Erb's phenomenon, 71  
   sign in tetany, 757  
   syphilitic paraplegia, 246, 828
- Erythroptosis, 480
- Exaltation, pathological, 60
- Excitability, affective, pathological, 62
- Exophthalmos in ocular paralysis, 485
- Extracerebellar tumors, 338
- Extradural hemorrhage, 257
- Eyes, conjugate deviation of, in lesion of internal capsule, 277  
   in hysteria, 684
- Eye-grounds in abscess of brain, 380  
   in tumor of brain, 328
- F**
- FACIAL atrophy in migraine, 719  
   hemiatrophy, 991  
   nerve, paralysis of, 495  
     in tabes dorsalis, 841  
   neuralgia, 733  
   paralysis, 495  
     course of, 501  
     diagnosis of, 501  
     etiology of, 495  
     fever in, 498  
     in hysteria, 679  
     meningitis and, 497  
     onset of, 498  
     otitis and, 497  
     pain in, 498  
     poliomyelitis and, 497  
     polyneuritis and, 497  
     prognosis of, 501  
     secondary contractures in, 502  
     symptoms of, 498  
     treatment of, 502  
       surgical, 503  
   spasm, 504  
     etiology of, 504  
     treatment of, 505
- Facio-scapulo-humeral progressive muscular dystrophy, 122
- Falling sickness, 592
- Family periodic paralysis, 809
- Fatigue, sense of, 19, 32
- Feelings, instability of, pathological, 62
- Femoral neuralgia, 740
- Fetal rickets, 955
- Fever in abscess of brain, 380  
   in apoplexy, 437  
   in arthritis deformans, 909, 914

Fever in caries of spine, 192  
in chorea, 779  
in dermatomyositis, 877  
in facial paralysis, 498  
in hysteria, 685  
in sciatica, 563  
in subacute combined sclerosis, 129  
in tetany, 759  
Fibrillary twitchings, 79  
Fibroma of brain, 317  
Fibrositis, 943  
acute, 944  
etiology of, 944  
pathology of, 944  
symptoms of, 944  
treatment of, 945  
Fingers in arthritis deformans, 912  
in hypertrophic osteo-arthritis, 984  
Foot-jerk, 77  
Fourth ventricle, tumors of, 338  
Fracture, intra-uterine, 979  
Fragilitas ossium, 947, 980  
congenital, 978  
Freud's hypothesis of hysteria, 657  
Friedreich's ataxia, 130. *See* Ataxia,  
Friedreich's  
Fröhlich's syndrome, 341  
Frontal lobe, tumors of, 331

G

GAIT, anomalies of, 73  
in Friedreich's ataxia, 136  
in lesion of cerebellum, 286  
in multiple neuritis, 581  
in osteomalacia, 951  
in paralysis agitans, 770  
Ganglia of cerebral hemisphere, 278  
inflammation of, neuralgia and, 726  
Gangrene in cervical ribs, 556  
Gastric crises in tabes dorsalis, 841, 849  
disease, tetany and, 749  
nerves, paralysis of, 523  
Gelle's test, 24  
Geniculate ganglia, inflammation of, 505  
Gerlier's disease, 515  
Gigantism, achondroplasia and, 961  
Glandular enlargement in arthritis de-  
formans, 909, 915  
Glaucoma in trigeminal neuralgia, 734  
Glioma of brain, diffuse, 312  
ependymal, 311  
of mid-brain, 284  
Gliomatosis in syringomyelia, 213  
Gliosis in epilepsy, 599  
Glossio-labio-laryngeal paralysis, 113  
Glossopharyngeal nerve, diseases of, 516  
Gluteal nerves, paralysis of, 558  
Glycosuria in tumors of brain, 338, 342  
Goitre, exophthalmic, diagnosis, of, from  
neurasthenia, 634  
Gout, arthritis deformans and, 930  
migraine and, 720  
Granuloma of brain, infectious, 319  
Gumma of brain, 320

H

HABIT chorea, 784  
spasm, 784  
Hallucinations, 37, 39  
auditory, 39  
complex, 39  
gustatory, 40  
olfactory, 40  
tactile, 40  
of visceral sense, 40  
visual, 39  
Hands in achondroplasia, 962  
in arthritis deformans, 912  
in hypertrophic osteo-arthritis, 984  
Headache in abscess of brain, 379  
in acute bulbar myelitis, 368  
meningitis, 170  
in anemia of brain, 393  
in arteriosclerosis of brain, 396  
in brain tumor, 324, 338  
in cerebral aneurism, 399  
in cerebrospinal syphilis, 824  
in lesion of pons, 290  
in Ménière's disease, 513  
in migraine, 716  
in neurasthenia, 631  
in pachymeningitis interna, 165  
in recurrent oculomotor palsy, 488  
sick, 709  
Hearing, anomalies of, 33  
centre of, 271  
in hemiplegia, 468  
in hysteria, 684  
in lesion of quadrigeminal bodies, 282  
in neurasthenia, 632, 634  
sense of, 22  
examination of, 23  
Heart disease, congenital, convulsions,  
and, 794  
Heberden's nodes in arthritis deformans,  
921  
diagnosis of, from hypertrophic  
osteo-arthritis, 987  
Hematomyelia, 260  
in cervical enlargement, 262  
in conus medullaris, 262  
diagnosis of, 263  
from acute poliomyelitis, 263  
from hematorrachis, 263  
from myelitis, 263  
from syringomyelia, 263  
in dorsal cord, 262  
etiology of, 260  
histology of, 261  
incontinence of urine in, 262  
in lumbosacral enlargement, 262  
onset of, 261  
paralysis in, 261  
pathology of, 261  
prognosis of, 263  
symptoms of, 261  
treatment of, 264  
Hematorrachis, 258

- Hematuria in polymyositis hemorrhagica, 879
- Hemeralopia, 35, 480
- Hemichromatopsia, 34  
in brain tumor, 334
- Hemiambyopia, 34
- Hemianesthesia, diagnosis of, from  
hysteria, 693  
in hysteria, 681  
in lesion of internal capsule, 276  
of pons, 290
- Hemianopic pupillary reaction, 482
- Hemianopsia, 34, 270, 481  
in abscess of brain, 381, 382, 384  
in brain tumor, 332, 334  
in hemiplegia, 468  
heteronyma, 34  
homonyma, 34  
in lesion of internal capsule, 276  
quadrantic, 34  
symptoms of, 271
- Hemiataxia in abscess of brain, 381, 384
- Hemiathetosis, 70  
in lesion of internal capsule, 277
- Hemiatrophy, facial, 991  
bones in, 992  
diagnosis of, 992  
hair in, 992  
prognosis of, 992  
skin in, 992  
symptoms of, 991  
treatment of, 992
- Hemicontracture in lesion of internal capsule, 277
- Hemicrania, 709
- Hemihyperesthesia in hysteria, 681
- Hemiparesis in abscess of brain, 384
- Hemiparesthesia in migraine, 712
- Hemiplegia, apoplexy and, 412, 457  
associated movements in, 462  
bedsores in, 469  
bladder in, 467  
cerebral, congenital, 409  
contractures in, 464  
gait in, 464  
hearing in, 468  
hemianopsia in, 468  
hysteria and, 424, 678  
infantile, 412  
brain tumor and, 419  
congenital, 409  
convulsions and, 415  
diphtheria and, 417  
embolism and, 418  
encephalitis and, 416  
etiology of, 413  
infections and, 417  
measles and, 417  
postnatal, 412  
scarlet fever and, 417  
syphilis and, 418  
typhoid fever and, 417  
whooping cough and, 417  
larynx in, 459  
in lesion of internal capsule, 276  
of optic thalamus, 279  
of pons, 290
- Hemiplegia in lesion of pontomedullocerebellar space, 290  
mastication in, 459  
mouth in, 460  
muscles in, 457, 461  
muscular rigidity in, 463  
tension in, 463  
palate in, 459  
posthemiplegic chorea in, 466  
ptosis in, 459  
reflexes in, deep, 465  
respiration in, 460  
sensation in, 467  
sympathetic effects in, 468  
tongue in, 460  
treatment of, 469  
tremor in, 465  
vasomotor effects in, 468
- Hemorrhage, apoplexy and, 422, 424, 427, 428, 431  
cerebral, 401, 410, 411, 414, 415  
consciousness in, 434  
onset of, 431  
extradural, 257  
intradural, 258  
diagnosis of, 259  
from hematomyelia, 263  
pain in, 258  
paralysis in, 258  
prognosis of, 259  
sensation in, 258  
symptoms of, 258  
treatment of, 260  
into spinal cord, 260  
membranes, 257
- Hereditary cerebellar ataxia, Marie's, 130, 136  
spastic spinal paralysis, 105
- Hernia, foraminal, of medulla, 324
- Herpes zoster, 571  
of auricle, 505  
in chorea, 781  
definition of, 571  
etiology of, 571  
fever in, 572  
hyperalgesia in, 572  
in migraine, 719  
pain in, 572, 573  
pathology of, 572  
skin in, 572  
symptoms of, 572  
treatment of, 573
- Hip disease, diagnosis of, from sciatica, 565
- Hip-joint in arthritis deformans, 913, 922
- Hoffmann's symptom, 71
- Hunger, sense of, 19, 31
- Huntington's chorea, 787  
diagnosis of, from Friedreich's ataxia, 139
- Hydatid cysts of brain, 319
- Hydrocephalus, 351  
acquired, 356  
cerebrospinal fever and, 357  
diagnosis of, 355  
etiology of, 356  
inflammation and, 357, 358

- Hydrocephalus, acquired, meningitis, posterior basic and, 357  
     serosa and, 358  
     tuberculous and, 357  
     tumor and, 356  
     agnosis of, from brain tumor, 345  
     epilepsy and, 599  
     essential, 353  
         cephalocele and, 354  
         course of, 354  
         diagnosis of, 355  
         from hydrocephalus, 355  
         from rickets, 355  
         etiology of, 353  
         pathology of, 354  
         spina bifida and, 354  
         symptoms of, 354  
         treatment of, 356  
     externus, 353  
     idiopathic, 353  
     internus, 353  
     secondary, 356  
     in tumor of brain, 338  
Hydromyelia, 210  
Hypacusis, 33  
Hypalgesia in tabes dorsalis, 838  
Hyperalgesia, 30, 33  
    in diseases of peripheral nerves, 539  
    in herpes zoster, 572  
    hysteria and, 682  
Hyperchondroplasia, 969  
Hyperemia of brain, 392  
    of spinal cord, 257  
Hyperesthesia, 28  
    in acute meningitis, 170  
    in hysteria, 681  
    of larynx, 522  
    in occupation neuroses, 747  
    in tabes dorsalis, 836  
Hypermetropia, 36  
Hypermnnesia, 53  
Hyperopia, 36  
Hyperostosis, 969  
    cranii, diagnosis of, from Paget's disease, 977  
    of skeleton with cyst-building, 978  
    of skull, general, 988  
        nodular, 989  
Hyperthymia, 60  
Hypertonicity in disseminated sclerosis, 147  
Hypertony of muscles, 68  
Hypertrophic internal pachymeningitis, 166  
    osteopathies, 969  
Hypertrophy of muscles, 68  
Hypesthesia in diseases of spinal roots, 568  
    in tabes dorsalis, 838  
Hypochondria, diagnosis of, from neurasthenia, 633  
Hypogastric reflexes, 78  
Hypoglossal nerve, diseases of, 530  
Hypomnesia, 53  
Hypopituitarism in brain tumor, 342  
Hypothymia, 61  
Hypotonia in lesion of cerebellum, 287  
    in tabes dorsalis, 839  
Hypotony of muscles, 68  
Hysteria, 654  
    adductor paralysis and, 521  
    akinetie motor phenomena in, 678  
    analgesia in, 682  
    anesthesia in, 680, 681  
    anuria in, 687  
    arhythmic movements in, 677  
    arthralgia in, 683  
    astasia-abasia and, 680  
    ataxia in, 652  
    attacks in, 671  
        varieties of, 675  
    Babinski's hypothesis of, 658  
    biological hypothesis of, 665  
    bleeding in, 687  
    blindness in, 684  
    blood in, 688  
    cardialgia in, 683  
    character in, 668  
    choreiform movements in, 677  
    contractures in, 651  
    convulsions in, 673  
    coördinated movements in, 676  
    course of, 675, 690  
    deep sensibility in, 682  
    dementia præcox and, 695  
    diagnosis of, 691  
        from acute meningitis, 172, 692  
        from apoplexy, 443  
        from brain tumor, 342  
        from chorea, 693  
        from dementia præcox, 694  
        from diseases of brain, 692  
        from disseminated sclerosis, 155  
        from encephalitis, 375  
        from epilepsy, 614, 691  
        from hemi-anesthesia, 693  
        from lateral sclerosis, 104  
        from manic-depressive insanity, 694  
        from multiple sclerosis, 692  
        from myasthenia gravis, 806  
        from neuralgia, 694  
        from neurasthenia, 633, 693  
        from organic disease, 650  
        from paresis, 694  
        from syringomyelia, 693  
    diarrhœa in, 688  
    dissociation in, 656  
    dizziness in, 684  
    dream states in, 674  
    emotional instability in, 669  
    emotions and, 698  
    enteralgia in, 683  
    etiology of, 654  
    eyes in, 684  
    facial paralysis in, 679  
    fever in, 685  
    Freud's hypothesis of, 657  
    glossolabial paralysis in, 679  
    hearing in, 684  
    hemi-anesthesia in, 681

Hysteria, hemihyperesthesia in, 681  
 hemiplegia and, 419, 424, 678  
 humoralism and, 654  
 hyperalgesia in, 682  
 hypesthesia in, 681  
 hypnotism in, 702  
 metabolism in, 688  
 monoplegia in, 681  
 motor disturbances in, 671  
 mutism in, 652  
 myasthenia in, 678  
 myoclonus in, 677  
 mythomania and, 659  
 negativism in, 670  
 nerve theories of, 655  
 neuralgia in, 683  
 cedema in, 686  
 ophthalmoplegia in, 679  
 paralysis in, 650, 678  
     of larynx in, 679  
 paraplegia in, 679  
 physiological theories of, 660  
 pithiatism and, 659  
 postconvulsive period of, 674  
 prognosis of, 690  
 pseudo-tabes and, 680  
 psychological hypothesis of, 655  
 psychopathology of, 654  
 psychoses in, 689  
 psychotherapy and, 702  
 ptosis in, 679  
 reëducation in, 704  
 reflexes in, 688  
 rhythmic movements in, 676  
 secretory disturbances in, 687  
 sensation in, 681  
 sensationalism in, 670  
 sight in, 684  
 simulation and, 659, 667  
 skin in, 686  
 smell in, 684  
 stigmata of, 667  
 suggestion in, 669, 697, 702  
 sympathetic system in, 663  
 symptoms of, 666  
 taste in, 684  
 temperature sense in, 682  
 tetany and, 677  
 topalgia in, 683  
 trauma and, 657  
 traumatic, 644, 691  
 treatment of, 696, 701  
     analytical or cathartic, 706  
     etiological, 700  
 tremors in, 676  
 trophic disturbances in, 685  
 vasomotor disturbances in, 685  
     hypothesis of, 662  
 vestibular apparatus in, 684  
 visceral pains in, 663  
 vision in, 652  
 Hysterical astasia-abasia, 815  
     disseminated sclerosis, 154  
     insanity, trauma and, 647  
     neuralgia, diagnosis of, 730  
     pseudo-tetany, diagnosis of, from  
     tetany, 760

## I

IDEAS, associative, anomalies of, 51  
     autochthonous, 52  
     duration of, anomalies of, 52  
     exaggerated, 52  
     falsification of content of, 56  
     flight of, 46  
     hyperdynamic, 52  
     imperative, 52  
         anomalies of conduct due to, 67  
         intensity of, anomalies of, 52  
 Ideation, anomalies of, 46  
 Idiocy, amaurotic family, 863  
     blindness in, 869  
     brain in, 864  
     diagnosis of, 869  
         from congenital diplegia  
         869  
         paraplegia, 869  
         from syphilis of brain,  
         870  
     etiology of, 864  
     fundus in, 863, 869  
     histology of, 866  
     pathogenesis of, 867  
     pathology of, 864  
     race and, 864  
     reflexes in, 869  
     symptoms of, 868  
     treatment of, 870  
 Idiopathic abscess of brain, 385  
 Illusions, 37, 38  
     memory, 55  
 Imbecility, anomalies of conduct due to,  
     67  
     epilepsy and, 594  
 Incoherence, 43, 51  
 Incoördination in syringomyelia, 221  
 Infantilism, 967  
     diagnosis of, from achondroplasia, 967  
 Infectious arthritis, 896  
 Infective myelitis, 229  
 Inflammation of brain, 359  
     of geniculate ganglia, 505  
     of optic nerve, 476  
 Influenza, arthritis deformans and, 901  
     neurasthenia and, 629  
 Influenzal encephalitis, 362  
 Insanity, diagnosis of, from neurasthenia,  
     633  
     epileptic, trauma and, 647  
     hysterical, trauma and, 647  
     manic-depressive, diagnosis of, from  
     hysteria, 694  
 Insular sclerosis, 140  
 Intercostal neuralgia, 739  
 Intermittent claudication, diagnosis of,  
     from neuralgia, 730  
     from sciatica, 565  
     disseminated sclerosis, 154  
 Interstitial hypertrophic progressive neu-  
     ritis of childhood, 112  
 Intestinal crises in tabes dorsalis, 842  
     disease, tetany and, 749  
     infection, arthritis deformans and,  
     903

Intracerebellar tumors, 338  
 Intracranial pressure, 391  
 Intradural hemorrhage, 258  
 Intra-ocular muscles, paralysis of, 486  
 Intra-uterine fracture, 979  
     rickets, 955  
 Irritability, pathological, 61  
 Irritation, motor, 70

**J**

JACKSONIAN epilepsy, 592  
 Joints in syringomyelia, 225

**K**

KERNIG'S sign in acute meningitis, 170  
 Kinanesthesia, 30  
 Klumpke palsy, 552  
 Knee-jerk, 77  
     in tabes dorsalis, 837  
 Knee-joint in arthritis deformans, 913,  
     923  
 Korsakow's syndrome, 45

**L**

LABYRINTH, choked, in brain tumors, 328  
 Landry's paralysis, 247. *See* Paralysis,  
     Landry's  
 Language, zone of, 293  
 Laryngeal muscles, spasm of, 522  
     nerves, paralysis of, 518  
         adductor, 521  
         diagnosis of, 519  
         larynx in, 519  
         phonation in, 519  
         recurrent, bilateral, 519  
         total unilateral, 519  
         respiration in, 519  
         treatment of, 520  
 Larynx in disease of vagus nerve, 517  
     in hemiplegia, 459  
     in myasthenia gravis, 804  
     sensory neuroses of, 522  
 Lasègue's sign in sciatica, 563  
 Lateral sclerosis, 101. *See* Sclerosis,  
     lateral.  
 Lathyrism, 139  
 Lead poisoning amyotrophic lateral scler-  
     osis and, 89  
 Lenticular degeneration, 278  
     nucleus, lesion of, 278  
     zone, lesion of, 278  
 Leontiasis ossea, 988  
 Leprosy, diagnosis of, from progressive  
     spinal muscular atrophy, 86  
 Leptomeninges in syphilitic myelitis, 239  
 Leptomeningitis, 167  
     acute, 167  
     chronic, 173  
     primary, 167  
     secondary, 168  
 Leukemia, retinitis in, 475

VOL. V—64

Leukocytosis in acute meningitis, 171  
     in arthritis deformans, 916  
 Lid reflex, 78  
 Lipoma of brain, 317  
 Lipomatosis, multiple, 818  
 Little's disease, 419  
 Liver, cirrhosis of, hypertrophic osteo-  
     arthropathy and, 982  
     in lenticular degeneration, 278  
 Lobstein's disease, 980  
 Locomotor ataxia, 833. *See* Tabes  
     dorsalis, 833  
     system, diseases of, 871  
 Logorrhœa, 46  
     in aphasia, 297  
 Lumbago, 945  
     traumatic, 644  
 Lumbar plexus, neuralgia of, 739  
     paralysis of, 560  
     puncture, 174  
         in acute meningitis, 171  
         in diagnosis of brain tumor, 330  
 Lumbo-abdominal neuralgia, 739  
 Lymphatics in adiposis dolorosa, 819  
 Lymphocytosis in spinal fluid, 177  
 Lymphorrhagia in myasthenia gravis, 799

**M**

MACROCEPHALY in achondroplasia, 963  
 Malaria, diagnosis of, from apoplexy,  
     444  
     migraines and, 720  
 Malnutrition, multiple neuritis and, 576  
 Mammary neuralgia, 739  
 Mania, acute, traumatism and, 647  
 Marie's hereditary cerebellar ataxia, 130,  
     136  
     theories in aphasia, 300  
 Mastication in hemiplegia, 459  
 Mastodynia, 739  
 Measles, apoplexy and, 417  
 Median nerve, paralysis of, 548  
 Medulla, hemorrhage of, diagnosis of, from  
     progressive bulbar palsy, 117  
     oblongata, foraminal hernia of, 324  
     tumors of, 337  
         diagnosis of, from progres-  
         sive bulbar palsy, 117  
 Megalocephalia, 988  
 Megalopsia, 480  
 Megrim, 709  
 Memory, anomalies of, 53  
     illusions, 55  
     phantasms, 55  
 Mendel's sign, 78  
 Ménière's disease, 512  
     diagnosis of, 513  
     pathology of, 513  
     prognosis of, 514  
     treatment of, 514  
     syndrome in brain tumor, 337  
 Meninges, anatomy of, 162  
     in cerebrospinal syphilis, 824  
     diseases of, 162  
     physiology of, 162

- Meningism, diagnosis of, from acute meningitis, 171
- Meningitis, blastomycotic, diagnosis of, from brain tumor, 345
- cerebrospinal, acute, 167
- Cheyne-Stokes respiration in, 170
- convulsions in, 170
- delirium in, 170
- diagnosis of, 171
- from brain abscess, 172
- from hysteria, 172
- from meningitis, 171, 172
- from otitis, 172
- etiology of, 167
- headache in, 170
- hyperesthesia in, 170
- influenza and, 168
- Kernig's sign in, 170
- leukocytosis in, 171
- lumbar puncture in, 171
- optic neuritis in, 170
- pain in, 170
- pathology of, 169
- photophobia in, 170
- pneumonia and, 168
- prognosis of, 173
- pulse in, 170
- reflexes in, 171
- rigidity of neck in, 170
- scarlet fever and, 168
- symptoms of, 169
- treatment of, 173
- vomiting in, 170
- diagnosis of, from abscess of brain, 386
- from hysteria, 692
- epilepsy and, 599
- facial paralysis and, 497
- localized serous spinal, 174
- posterior basic, hydrocephalus and, 357
- serosa, hydrocephalus and, 358
- serous, diagnosis of, from brain tumor, 345
- tuberculous, diagnosis of, from acute otitis, 172
- hydrocephalus and, 357
- Meningo-encephalitis, tuberculous, of brain, 319
- Meningomyelitis, syphilitic, diagnosis of, from spinal caries, 193
- from tabes dorsalis, 845
- Menopause, arthritis deformans and, 902
- Mental disturbances, traumatic, 646
- failure in amyotrophic lateral sclerosis, 99
- fog, 44
- phenomena in tetany, 758
- Meralgia paresthetica, 557
- anterior, 558
- Metabolism in arthritis deformans, 903, 907
- in hysteria, 688
- multiple neuritis and, 575
- osteomalacia and, 949, 952
- Metacarpophalangeal joints in arthritis deformans, 922
- Metastatic abscess of brain, 384
- Microcephalia, 990
- Micromelie rickets, 955
- Micturition in disseminated sclerosis, 150
- Mid-brain, glioma of, 284
- lesion of, 282
- ataxia in, 283
- paralysis in, 283
- Migraine, ophthalmic, 709
- anesthesia in, 713, 714
- abortive, 711
- agraphia in, 715
- cerebral tumor and, 720
- chilliness in, 713
- classical, 713
- combined hypothesis of, 710
- definition of, 709
- diagnosis of, 720
- dizziness in, 716
- epilepsy and, 615, 711
- etiology of, 710
- facial atrophy in, 719
- gout and, 720
- headache in, 716
- hemiparesthesia in, 712
- heredity and, 711
- herpes zoster in, 719
- malarial fever and, 720
- motor disturbances in, 715
- pain in, 717
- paralytic phenomena in, 716
- paresthesia in, 713, 714
- paresis and, 720
- psychical disturbances in, 719
- reflex hypothesis of, 710
- scotomata in, 712, 714
- secretions in, 718
- speech in, 715
- status hemicanianus in, 718
- symptomatic, 720
- symptoms of, cerebellar, 716
- early, 713
- sensory, 713
- visual, 712, 714
- tabes and, 720
- theories of, 710
- toxic hypothesis of, 710
- treatment of, 721
- trophic disturbances in, 718
- vasomotor disturbances in, 718
- hypothesis of, 710
- vomiting in, 712
- ophthalmoplegic, 724
- diagnosis of, 725
- etiology of, 724
- symptoms of, 725
- treatment of, 725
- periodic paralysis and, 811
- Miliary sclerosis, 161
- Millard-Gubler paralysis, 290
- in acute encephalomyelitis, 369
- Mind blindness, 53, 271, 483
- in aphasia, 299
- deafness, 54
- Mollities ossium, 947



- Molluscum fibrosum, 571  
 Monarticular arthritis deformans, 919, 934  
 Monocular diplopia, 480  
 Monoideic somnambulism, 44  
 Monoplegia in hysteria, 681  
 Moods, 59, 62  
 Morvan's type of syringomyelia, 220  
 Motility, disturbances of, 68  
 Motor actions, acceleration of, 64  
     incoherence of, 66  
     retardation of, 65  
     agitation, 63  
     aphasia, 75, 292, 295  
     area of cortex of brain, 265  
     disturbances in migraine, 715  
         in neuralgia, 728  
     irritation, 70  
     nerves in tetany, 757  
     perseveration, 67  
     phenomena, in infantile convulsions, 794  
     stupor, 63, 65  
 Mouth in hemiplegia, 460  
 Movement, sensation of, 17  
 Mucous membrane in hysteria, 682  
 Multiple neuritis, 573  
     sclerosis, 140  
 Muscles in amyotonia congenita, 894  
     in arthritis deformans, 907  
     atony of, 68  
     atrophy of, 68  
         in arthritis deformans, 914  
         in diseases of peripheral nerves, 534  
         in paralysis agitans, 771  
         progressive spinal, 82  
     contractures of, 68  
     cramps of, in tetany, 754  
     disturbed innervation of, in lesion of optic thalamus, 280  
     electrical reactions of, 72  
     hypertony of, 68  
     hypertrophy of, 68  
     hypotony of, 68  
     inflammation of, 871. *See* Myositis.  
     irritability of, in tetany, 758  
     irritation of, 70  
     in myasthenia gravis, 799, 802  
     in myotonia congenita, 887  
     of neck, spasm of, 525  
     ocular, paralysis of, 485  
     paralyses of, 68  
     in paramyoclonus multiplex, 807  
     in periodic paralysis, 813  
     in progressive myositis ossificans, 883  
     rigidity of, in paralysis agitans, 776  
     sense, 17  
     spasms of, 71  
     tension of, in diseases of peripheral nerves, 534  
         in hemiplegia, 463  
     of tongue, spasm of, 532  
     tonus of, 68  
 Muscular atrophy, 80  
     dystrophy, diagnosis of, from myasthenia gravis, 806  
 Muscular dystrophy, progressive, 117  
     hypotonia in tabes dorsalis, 839  
     rigidity in hemiplegia, 463  
         in paralysis agitans, 769  
     wasting in spondylitis deformans, 927  
     weakness in general paresis, 857  
 Musculocutaneous nerve, paralysis of, 546  
 Musculospiral nerve, paralysis of, 547  
 Mutism in hysteria, 652  
 Myalgia, diagnosis of, from neuralgia, 730  
 Myasthenia gravis, 798  
     age and, 798  
     bulbar muscles in, 802  
     diagnosis of, 805  
         from astasia-abasia, 816  
         from diphtheritic paralysis, 806  
         from hysteria, 806  
         from muscular dystrophy, 806  
         from policephalitis, 806  
         from progressive bulbar palsy, 116, 806  
         muscular dystrophy, 124  
     diplopia in, 802  
     dyspnea in, 804  
     etiology of, 798  
     extremities in, 804  
     in hysteria, 678  
     infections and, 798  
     larynx in, 804  
     lymphorrhages in, 799  
     muscles in, 799, 802  
     myasthenic reaction in, 801, 805  
     nervous diseases and, 798  
     pathology of, 799  
     pregnancy and, 799  
     prognosis of, 806  
     reflexes in, 805  
     sex and, 798  
     symptoms of, 802  
     thymus gland and, 799  
     toxins and, 800  
     treatment of, 807  
     urine in, 805  
     von Graefe's sign in, 803  
 Myasthenic reaction, 801, 805  
 Mydriasis, 77  
     in tabes dorsalis, 837  
 Myelitis, 227  
     acute ascending, 236  
         bulbar, 368  
     bacteria and, 229  
     chronic, 245  
     compression, 229  
     definition of, 227  
     degenerative, 236  
     diagnosis of, 241  
         from disseminated sclerosis, 155  
         from hematomyelia, 263  
         from Landry's paralysis, 252  
         from lateral sclerosis, 104  
         from peripheral neuritis, 242  
     disseminated, 235  
     infective, 229  
         anesthesia in, 234  
         bacteriology of, 231

- Myelitis infective, cervical, 234  
 course of, 235  
 dorsal, 234  
 etiology of, 229  
 histology of, 232  
 lumbosacral, 235  
 nervous elements in, 233  
 neuroglia in, 232  
 pathogenesis of, 229  
 pathology of, 231  
 symptoms of, 233  
 transverse, 234  
 treatment, 243  
 vascular changes in, 232  
 prognosis of, 243  
 syphilis and, 228  
 syphilitic, 237, 828  
 age and, 237  
 alcoholism and, 238  
 bladder in, 240  
 bloodvessels in, 238  
 course of, 241  
 diagnosis of, 242, 828  
 etiology of, 237  
 histology of, 238  
 leptomeninges in, 239  
 nervous elements in, 239  
 neuroglia in, 239  
 onset of, 240  
 pain in, 240  
 paralysis in, 240  
 paraplegia in, 241  
 pathology of, 238, 828  
 sex and, 237  
 symptoms of, 240, 828  
 treatment of, 244, 830  
 toxic, 236  
 differentiation from infective, 236  
 etiology of, 236  
 pathology of, 237  
 pregnancy and, 236  
 symptoms of, 237  
 treatment of, 243  
 treatment of, 244  
 Myeloma of brain, 315  
 of spinal cord, 198  
 Myeloplastic bone aplasia, 978  
 Myoclonus in hysteria, 677  
 Myopathy, primary, 117  
 Myopia, 36  
 Myopsia, 480  
 Myosis, 77  
 Myositis, 871  
 acute, diagnosis of, from neuritis, 587  
 definition of, 871  
 non-suppurative, 875  
 ossificans, progressive, 881  
 bones in, 883, 884  
 diagnosis of, 885  
 from locomotor ataxia, 885  
 from multiple exostoses, 885  
 from spondylitis deformans, 885  
 etiology of, 881  
 fever in, 883  
 Myositis ossificans, progressive, muscles  
 in, 883  
 œdema in, 883  
 pain in, 883  
 pathology of, 882  
 prognosis of, 885  
 symptoms of, 883  
 treatment of, 885  
 primary, fibrosa, 880  
 suppurative, 872  
 chills in, 873  
 diagnosis of, 874  
 from metastatic abscesses, 874  
 from osteomyelitis, 874  
 ecchymosis in, 874  
 erythema in, 874  
 etiology of, 872  
 fever in, 873  
 œdema in, 874  
 pain in, 873  
 pathology of, 873  
 prognosis of, 874  
 symptoms of, 873  
 treatment of, 875  
 with special terminal lesions, 880  
 Myotonia congenita, 886  
 age and, 886  
 cramps in, 890  
 diagnosis of, 892  
 electrical reactions in, 891  
 etiology of, 886  
 muscles in, 887, 890  
 myotonic reaction in, 891  
 pathogenesis of, 889  
 pathology of, 887  
 prognosis of, 893  
 reflexes in, 892  
 sex and, 886  
 symptoms of, 890  
 treatment of, 893  
 Myotonic reaction, 891  
 Mythomania, 659  
 Myxosarcoma of brain, 315
- ## N
- NAILS in diseases of peripheral nerves, 541  
 in osteo-arthritis, 984  
 Neck, muscles of, spasm of, 525  
 rigidity of, in acute meningitis, 170  
 Nephritis, chronic, apoplexy and, 427  
 diagnosis of, from brain tumor, 344  
 in polymyositis hemorrhagica, 880  
 Nerve deafness, 509  
 Rinné's test in, 510  
 Weber's test in, 510  
 irritability of, in tetany, 758  
 motor, in tetany, 757  
 section of, recovery from, 538  
 sensation and, 536  
 sensory, irritation of, 539  
 trunks, neuralgia and, 726  
 tumors of, 568. *See* Neuromata.  
 Nervous system, arthritis deformans and, 903, 907

- Nervous system in dermatomyositis, 877  
 myasthenia gravis and, 798  
 in Paget's disease, 973  
 parasymphilis of, 822  
 syphilis of, 822  
   spirochæta pallida in, 823  
 tetany and, 749
- Neuralgia, 725  
 anemia and, 726  
 anesthesia in, 728  
 arteriosclerosis and, 727  
 brachial, 737  
   diagnosis of, 738  
   pain in, 738  
   symptoms of, 738  
   treatment of, 738  
 cervico-occipital, 736  
   etiology of, 736  
   pain in, 736  
   Valleix's points in, 737  
 of coccygeal plexus, 741  
 counterirritation in, 732  
 course of, 729  
 crural, 740  
 definition of, 725  
 diagnosis of, 729  
   from hysteria, 694  
   from intermittent claudication, 730  
   from myalgia, 730  
   from neurasthenia, 730  
   from neuritis, 730  
   from tabes dorsalis, 730  
 diaphragmatic, 737  
 etiology of, 726  
 facial, 733  
 femoral, 740  
 ganglia and, 726  
 in hysteria, 683  
 hysterical, diagnosis of, 730  
 intercostal, 739  
 localized forms of, 733  
 of lumbar plexus, 739  
 lumbo-abdominal, 739  
 mammary, 739  
 motor disturbances in, 728  
 nerve trunks and, 726  
 obturator, 740  
 pain in, 727  
 paresis and, 727  
 paresthesia in, 728  
 prognosis of, 731  
 psychological, 726  
 of pudendal plexus, 740  
 reflex, 726, 731  
 sciatic, diagnosis of, from simple sciatica, 565  
 secretory disturbances in, 728  
 skin in, 728  
 symptoms of, 727  
 syphilis and, 727  
 tabes dorsalis and, 727  
 testicular, 740  
 toxins and, 726  
 treatment of, 731  
   surgical, 732  
 trigeminal, 733
- Neuralgia, trigeminal, course of, 735  
 diagnosis of, 735  
 exposure and, 733  
 glaucoma in, 734  
 in multiple sclerosis, 733  
 pain in, 734  
 photophobia in, 734  
 skin in, 734  
 symptoms of, 733  
 in tabes dorsalis, 733  
 tic douloureux, 733  
 treatment of, 735  
 Valleix's points in, 734  
 visual disturbances in, 734  
 Valleix's points in, 727  
 vasomotor disturbances in, 728
- Neurasthenia, 624  
 appetite in, 631  
 in arteriosclerosis of brain, 395  
 circulation in, 631  
 climate and, 628  
 diagnosis of, 632  
   from Graves' disease, 634  
   from hypochondria, 633  
   from hysteria, 633, 693  
   from insanity, 633  
   from neuralgia, 730  
 emotional stress and, 627, 630  
 etiology of, 624  
 headache in, 631, 634  
 hearing in, 632, 634  
 heredity and, 625  
 influenza and, 629  
 isolation in, 637  
 occupation and, 627  
 pain in, 631  
 paresthesia in, 631  
 prognosis of, 634  
 psychotherapy in, 640  
 race and, 628  
 reflexes in, 632  
 secondary, 628  
 symptomatic, 628  
 symptoms of, 630  
 syphilis and, 629  
 in tabes dorsalis, 843  
 traumatic, 643  
 treatment of, 635  
 typhoid fever and, 629  
 urine in, 632  
 vertigo in, 631  
 vision in, 632
- Neuritis, brachial, 553  
 diagnosis of, from neuralgia, 730  
 interstitial hypertrophic progressive, of childhood, 112  
 multiple, 573  
   alcoholic, 579  
     diagnosis of, from tabes dorsalis, 845  
   anemia in, 585  
   ascending, 576  
   ataxia in, 583  
   bacteria and, 575, 576  
   cachexia and, 576  
   classification of, 574  
   complications of, 586

- Neuritis, multiple, course of, 585  
   cranial nerves in, 582  
   diagnosis of, 586  
     from acute myositis, 587  
       poliomyelitis, 586  
     from Landry's paralysis, 586  
     from cedema, 587  
     from progressive bulbar palsy, 116  
   neural, muscular atrophy, 111  
   spinal muscular atrophy 86  
     from tabes dorsalis, 587  
     from trichiniasis, 587  
   diphtheria and, 575  
   electrical reactions in, 582  
   etiology of, 574  
   gait in, 581  
   incidence of, 576  
   malnutrition and, 576  
   metabolism and, 575  
   cedema in, 585  
   onset of, 579  
   paralysis in, 580  
   pathology of, 576  
   poisons and, 575  
   polyarthritis and, 585  
   prognosis of, 588  
   reflexes in, 584  
   scarlet fever and, 575  
   smallpox and, 575  
   sphincters in, 584  
   symptoms of, 579  
     mental, 585  
     motor, 580  
     sensory, 583  
   tenderness in, 583  
   treatment of, 588  
   trophic changes in, 584  
   tuberculosis and, 575, 586  
   typhoid fever and, 575  
   vasomotor changes in, 584  
   optic, 36  
     in acute meningitis, 170  
   peripheral, diagnosis of, from syphilitic myelitis, 242  
   retrobulbar, 37, 479  
   sciatic, diagnosis of, from organic sciatica, 565  
 Neuroblastoma of brain, 317  
 Neurocytoma of brain, 317  
 Neurofibromatosis, general, 570  
 Neuroglia in infective myelitis, 232  
   in syphilitic myelitis, 239  
 Neuroglioma of brain, 311  
 Neuroma of brain, 317  
 Neuromata, 568  
   false, 569  
   multiple, 590  
   pathology of, 569  
   plexiform, 571  
   solitary, 569  
   true, 568  
 Neuromyositis, 880  
 Neuroses of larynx, sensory, 522  
   occupation, 741  
 Neuroses, occupation, age and, 743  
   anesthesia in, 747  
   atrophy in, 744  
   course of, 747  
   cramps in, 744  
   definition of, 741  
   etiology of, 742  
   heredity and, 742  
   hyperesthesia in, 747  
   occupation and, 743  
   occurrence of, 743  
   palsies in, 744  
   paresthesia in, 747  
   prognosis of, 747  
   psychical factors in, 742  
   sensory disturbances in, 747  
   symptoms of, 743  
   synonyms of, 741  
   treatment of, 747  
   writer's cramp, 744  
   professional, 741  
   traumatic, 642  
     anesthesia in, 651  
     ataxia in, 652  
     contractures in, 651  
     diagnosis of, 648  
     fraud in, 642  
     mutism in, 652  
     paralysis in, 651  
     prognosis of, 652  
     vision in, 652  
 Neuro-tabes peripherica, 579, 584  
 Neurotic progressive facial atrophy, 991  
 Night blindness, 35, 480  
 Nodosity of joints, 897  
 Nyctalopia, 35, 480  
 Nystagmus, 70  
   in abscess of brain, 384  
   in brain tumor, 336, 339, 340  
   in disseminated sclerosis, 151  
   in Friedreich's ataxia, 135  
   in Ménière's disease, 513  
   in ocular paralysis, 486  
   in pseudo-sclerosis, 159  
   in syringomyelia, 222  
   in vestibular disease, 511
- O
- OBLIQUE muscles, paralysis of, 485  
 Obsessions, 52  
 "Obstetrical hand," 71  
 Obturator nerve, paralysis of, 557  
 Occupation neuroses, 741  
 Ocular muscles in disseminated sclerosis, 151  
   paralysis of, 485  
     exophthalmos in, 485  
     nystagmus in, 486  
 nerves, paralysis of, 483  
   in meningitis, 488  
   ophthalmoplegia in, 483  
   ptosis in, 488  
   symptoms of, 483  
   in syphilis, 488  
   in tabes, 488

- Ocular palsy, nuclear, 365
- Oculomotor nerve, anatomy of, 483
- palsy, recurrent, 488
- in tabes dorsalis, 841
- Œdema in amyotonia congenita, 894
- in arthritis deformans, 917
- in brain tumor, 323
- in dermatomyositis, 877
- diagnosis of, from neuritis, 587
- in hysteria, 686
- in multiple neuritis, 585
- in polymyositis hemorrhagica, 879
- Œsophageal nerves, paralysis of, 523
- Olfactory nerve, diseases of, 471
- Olivopontocerebellar atrophy, 287
- Ophthalmoplegia, 483
- in acute encephalomyelitis, 369
- in cerebrospinal syphilis, 825
- chronic, 491
- external, 86
- diplopia in, 484
- externa, 490
- in hysteria, 678
- interna, 490
- in lesion of quadrigeminal bodies, 283
- strabismus in, 484
- in superior hemorrhagic poli-encephalitis, 365
- in tabes dorsalis, 841
- Ophthalmoplegic migraine, 724
- Ophthalmoscopic examination in apoplexy, 437
- Opium poisoning, diagnosis of, from apoplexy, 442
- Oppenheim's disease, 893
- sign, 78
- Optic aphasia, 54, 299
- asymbolia, 54
- atrophy, 478
- diagnosis of, 479
- primary, 479
- secondary, 479
- chiasm, affections of, 481
- disks, in disseminated sclerosis, 152
- in papillitis, 478
- disseminated sclerosis, 154
- nerve, anatomy of, 473
- atrophy of, 37
- in tabes dorsalis, 841
- diseases of, 476
- inflammation of, 476
- neuritis, 36, 326
- thalamus, lesion of, 279
- astereognosis in, 281
- ataxia in, 280
- athetosis in, 280
- disturbed innervation of
- muscles in, 280
- hemiplegia in, 279
- tracts, affections of, 481
- Osteitis, chronic parenchymatous, 978
- deformans, 969, 970
- arteriosclerosis in, 973
- bones in, 971
- deformity in, 974
- diagnosis of, 976
- from acromegaly, 977
- Osteitis deformans, diagnosis of, from hyperostosis cranii, 977
- from osteo-arthropathy, 977
- from osteomalacia, 977
- from spondylitis, 977
- etiology of, 970
- heredity and, 970
- occurrence of, 970
- pain in, 974, 975
- pathology of, 970
- prognosis of, 977
- skull in, 971
- spine in, 971
- symptoms of, 973
- syphilis and, 973
- treatment of, 977
- tumor-building, 978
- tumors and, 973
- fibrosa, 980
- Osteo-arthritis, 897, 920
- Osteo-arthropathy, hypertrophic pulmon-ary, 981
- age and, 983
- arthritis in, 985
- atrophy in, 986
- bones in, 984
- bronchiectasis and, 982
- cirrhosis of liver and, 982
- cyanosis and, 982
- diagnosis of, 987
- from arthritis defor-mans, 987
- from Heberden's nodes, 987
- from Paget's disease, 977
- etiology of, 981
- fingers in, 984
- hands in, 984
- nails in, 984
- pain in, 985
- pathology of, 983
- prognosis of, 987
- race and, 983
- sex and, 983
- symptoms of, 984
- syphilis and, 982
- treatment of, 988
- tuberculosis and, 981
- types of, 986
- secondary, 981
- Osteogenesis imperfecta, 978
- bones in, 979
- etiology of, 979
- intra-uterine fracture and, 979
- pathology of, 979
- Osteomalacia, 947
- age and, 947
- bones in, 949
- congenita, 978
- definition of, 947
- diagnosis of, 953
- from Paget's disease, 977
- diet in, 953
- etiology of, 947
- gait in, 951
- metabolism and, 949, 952

- Osteomalacia, ovaries and, 948  
 pain in, 951  
 parathyroid gland and, 949  
 pathology of, 948  
 pregnancy and, 948  
 reflexes in, 951  
 sex and, 947  
 symptoms of, 951  
 tetany and, 949  
 thyroid gland in, 948  
 treatment of, 953  
 urine in, 952  
 varieties of, 953  
 vertebræ in, 950  
 with cyst-building, 978
- Osteomyelitis, diagnosis of, from primary  
 suppurative myositis, 874
- Osteopathies, hypertrophic, 969
- Osteoporosis congenita, 978
- Osteopsathyrosis, 978, 980  
 etiology of, 980  
 prognosis of, 981  
 symptoms of, 981  
 treatment of, 981
- Osteosarcoma of brain, 314
- Osteosclerosis, congenital, 968, 978
- Ostitis deformans, 970  
 fibrosa hyperostotica cranii et faciei,  
 988
- Otitic abscess of brain, 383
- Otitis, diagnosis of, from acute meningitis,  
 172
- Ovaries in osteomalacia, 948

## P

- PACHYMENINGITIS, 163  
 cervicalis hypertrophica, 166  
 diagnosis of, 167  
     from progressive spinal  
     muscular atrophy, 86  
     from tumors of spinal  
     cord, 208  
 etiology of, 166  
 symptoms of, 166  
 treatment of, 167
- externa, 163  
 cerebral, 163  
 spinal, 163  
 symptoms of, 164  
 treatment of, 164
- interna, 164  
 aphasia in, 165  
 convulsions in, 165  
 diagnosis of, 165  
 headache in, 165  
 hemorrhagica, 164  
 optic neuritis in, 165  
 pathology of, 164  
 symptoms of, 165  
 treatment of, 166  
 vomiting in, 165
- Paget's disease, 970
- Pain in abscess of brain, 381, 384  
 in acute meningitis, 170  
 in adiposis dolorosa, 819, 820
- Pain in amyotrophic lateral sclerosis, 99  
 in arthritis deformans, 909, 910, 921,  
 922  
 in brachial neuritis, 553  
 in brain tumor, 338  
 in cervical ribs, 555  
 in cervico-occipital neuralgia, 736  
 in compression of spinal cord, 190  
 in dermatomyositis, 877  
 in diseases of spinal roots, 567  
 in disseminated sclerosis, 149  
 in facial paralysis, 498  
 in Friedreich's ataxia, 136  
 in herpes zoster, 572  
 in intercostal neuralgia, 739  
 in intradural hemorrhage, 258  
 in Landry's paralysis, 251  
 in lesion of optic thalamus, 280  
     of spinal cord, 183  
 in neuralgia, 727  
 in neurasthenia, 631  
 in ophthalmic migraine, 717  
 in osteo-arthropathy, 985  
 in pachymeningitis cervicalis hyper-  
 trophica, 166  
 in Paget's disease, 974, 975  
 in paralyzing vertigo, 515  
 in polymyositis hemorrhagica, 879  
 in primary myositis fibrosa, 881  
 in recurrent oculomotor palsy, 488  
 in sciatica, 562  
 sense, 17  
     anomalies of, 29  
 in solitary neuromata, 569  
 in spasmodic torticollis, 528  
 in spondylitis deformans, 925, 926  
 in syphilitic myelitis, 240  
 in tabes dorsalis, 836  
 in tetany, 757, 759  
 in trigeminal neuralgia, 734  
 in tumors of spinal cord, 201, 202
- Palate in hemiplegia, 459  
 paralysis of, in hysteria, 679
- Pallanesthesia, 30
- Palmar reflex, 78
- Palsies, cerebral, congenital, 410  
 Duchenne-Erb, 551  
 Klumpke's, 552  
 in occupation neuroses, 744  
 oculomotor, recurrent, 488  
     in tabes dorsalis, 841
- Palsy, bulbar, acute inflammatory, 368  
 of childhood, diagnosis of, from  
 progressive bulbar palsy, 117  
 diagnosis of, from progressive  
 bulbar palsy, 117  
 progressive, 113  
     diagnosis of, 116  
     from bulbar palsy, 117  
     of childhood,  
     117  
     type of muscular  
     dystrophy, 117  
     from encephalitis, 117  
     from hemorrhage, 117  
     from multiple neuritis,  
     116

- Palsy, bulbar, progressive, diagnosis of,  
     from myasthenia gravis, 116  
     from tumor of pons and medulla, 117  
     etiology of, 113  
     pathology of, 114  
     reflexes in, 116  
     speech in, 115  
     symptoms of, 115  
     treatment of, 117  
     shaking, 764
- Papillitis, 326  
     mechanism of, 476  
     optic disk in, 478
- Papilloma of brain, 315
- Paracentral convolutions, tumors of, 332
- Paragraphia, 66, 77
- Paralysis in abscess of brain, 381  
     in acute infective polyneuritis, 590
- agitans, 764  
     age and, 764  
     atrophy in, 771  
     definition of, 764  
     diagnosis of, 772  
         from chorea, 773  
         from multiple sclerosis, 772  
         from senile tremor, 773  
     ductless glands and, 767  
     etiology of, 764  
     face in, 770  
     forms of, 772  
     gait in, 770  
     heredity and, 764  
     parathyroid glands and, 767  
     pathogenesis of, 767  
     pathology of, 765  
     prognosis of, 773  
     reflexes in, 771  
     rigidity in, 769  
     symptoms of, 768  
     trauma and, 764  
     treatment of, 773  
     tremor in, 768  
     voice in, 771
- in amaurotic family idiocy, 869  
     in apoplexy, 438, 440  
     ascending bulbar, 798  
     asthenic, 798  
     atrophic, in syringomyelia, 221  
     birth, brachial, 552  
     of brachial plexus, 550  
     in brain tumor, 337  
     of cardiac nerves, 522  
     in cerebrospinal syphilis, 825  
     of cervical plexus, 543  
     of circumflex nerve, 546  
     diaphragmatic, 544  
     diphtheritic, diagnosis of, from myasthenia gravis, 806  
     Duchenne-Erb, 551  
     of external rectus muscle, 485  
     of facial nerve, 495  
     flaccid, 178  
     of gastric nerves, 523  
     glosso-labio-laryngeal, 113  
     of gluteal nerves, 558
- Paralysis in hematomyelia, 260  
     in hysteria, 650, 678, 679  
     diagnosis of, 650, 678  
         from apoplexy, 446  
     in infantile convulsions, 795  
     of inferior oblique muscle, 485  
         rectus muscle, 485  
     of internal rectus muscle, 485  
     in intradural hemorrhage, 258  
     of intra-ocular muscles, 486  
     Klumpke's, 552  
     Landry's, 247  
         bacteriology of, 248  
         bladder in, 251  
         diagnosis of, 252  
             from myelitis, 252  
             from neuritis, 586, 587  
             from poliomyelitis, 252  
     dyspnea in, 251  
     etiology of, 248  
     pain in, 251  
     paresthesia in, 251  
     pathology of, 249  
     prognosis of, 251  
     symptoms of, 250  
     treatment of, 254  
     vasomotor disturbances in, 251
- laryngeal, 518, 521
- in lesion of cerebellum, 286  
     of crura cerebri, 281  
     of internal capsule, 276  
     of mid-brain, 283  
     of pons, 289  
     of tegmentum, 281
- of long thoracic nerve, 544  
     of lower arm, 552  
     of lumbar plexus, 560  
     of median nerve, 548  
     Millard-Gubler type, 290  
     in multiple neuritis, 580  
     of muscles, 68  
     of musculocutaneous nerve, 546  
     of musculospiral nerve, 547  
     of nerves of legs, 556  
         diagnosis of, 560  
             from disease of cord, 560  
             from lesions of cauda equina, 561  
             of sacral plexus, 561
- of ocular nerves, 483  
     sympathetic, in syringomyelia, 224
- of oesophageal nerves, 523
- periodic, 809  
     auto-infection and, 811  
     blood in, 811  
     course of, 814  
     definition of, 809  
     etiology of, 810  
     heredity and, 810  
     migraine and, 811  
     muscles in, 813  
     paralysis in, 813  
     pathology of, 810  
     pulse in, 813  
     reflexes in, 814  
     symptoms of, 812  
     transmission of, 810

- Paralysis, periodic, treatment of, 814  
     urine in, 811  
     of pharyngeal nerves, 517  
     of phrenic nerve, 543  
     of popliteal nerves, 559  
     of posterior tibial nerve, 559  
     in progressive spinal muscular atrophy, 84  
     pseudo-bulbar, 290, 978  
     of pulmonary nerves, 523  
     of radial nerve, 547  
     of sacral plexus, 560  
     of sciatic nerve, 558, 560  
     "soul," 268  
     spastic, 179  
         spinal, 101  
             hereditary, 105  
             in syringomyelia, 221  
     of spinal accessory nerve, 524  
     of superior oblique muscle, 486  
         rectus muscle, 485  
     of suprascapular nerve, 545  
     in syphilitic myelitis, 240, 828  
     of tongue, 530  
     in traumatic neuroses, 651  
     of trigeminal nerve, 493, 494  
     of ulnar nerve, 549  
     unilateral, 108  
     of upper arm, 551  
     of vagus nerve, 517
- Paralytic conjugate deviation in apoplexy, 439  
     phenomena in chorea, 780  
     in migraine, 716
- Paramimia, 66
- Paramnesia, 55
- Paramyoclonus multiplex, 807  
     diagnosis of, 808  
     etiology of, 807  
     muscles in, 808  
     pathology of, 807  
     prognosis of, 809  
     symptoms of, 808  
     treatment of, 809
- Paranoia, trauma and, 647
- Paraphasia, 66  
     in abscess of brain, 381
- Paraplegia in cerebrospinal syphilis, 825  
     congenital, diagnosis of, from amaurotic family idiocy, 869  
     Erb's syphilitic, 246, 828  
     in hysteria, 679  
     spastic, diagnosis of, from disseminated sclerosis, 156  
     in subacute confined sclerosis, 127, 128  
     in syphilitic myelitis, 241  
     in tumors of spinal cord, 203
- Parapraxia, 66
- Parasitic cysts of brain, 318
- Parasyphilis of nervous system, 822
- Parathyroid gland, osteomalacia and, 949  
     tetany and, 750, 753
- Parenchymatous multiple neuritis, 577
- Paresis, general, 850  
     Argyll-Robertson pupil in, 856  
     convulsions in, 857  
     course of, 857
- Paresis, general, delusions in, 855  
     diagnosis of, 859  
         from alcoholic pseudo-paresis, 860  
         from arteriosclerosis, 398  
         from arteriosclerotic dementia, 861  
         from brain tumor, 343  
         from disseminated sclerosis, 156, 860  
         from syphilitic pseudo-paresis, 860  
     duration of, 858  
     etiology of, 850  
     hematoma in, 857  
     memory in, 854  
     pathology of, 852  
     prognosis of, 859  
     pupils in, 856  
     reflexes in, 857  
     speech in, 856  
     Spirochaeta pallida in, 851, 852  
     symptoms of, 853  
     syphilis and, 850  
     tabes dorsalis and, 843  
     trauma and, 647  
     treatment of, 861  
     tremors in, 856  
     trophic disturbances in, 858  
     vasomotor disturbances in, 858  
     Wassermann reaction in, 851  
     migraine and, 720  
     neuralgia and, 727
- Paresthesia in diseases of peripheral nerves, 539  
     of spinal roots, 567  
     in Landry's paralysis, 251  
     of larynx, 522  
     in migraine, 713, 714  
     in neuralgia, 728  
     in neurasthenia, 631  
     in occupation neuroses, 747  
     in osteomalacia, 951  
     in sciatica, 563  
     in solitary neuromata, 570
- Paretic gait, 73
- Parietal lobe, tumors of, 333
- Parkinson's disease, 764
- Patellar tendon reflex, 77
- Pellagra, 139
- Pelvis, tumor in, diagnosis of, from sciatica, 565
- Perception, anomalies of, 37  
     sense, 18
- Peri-articular arthritis deformans, 908
- Pericarditis in arthritis deformans, 914
- Periodic paralysis, 809
- Periosteal dyscrasia, 978
- Peripheral nerves, diseases of, 533  
     atrophy in, 534  
     causalgia in, 539  
     deep reflexes in, 534  
     distal reaction in, 535  
     electrical reactions in, 535  
     hyperalgesia in, 539  
     loss of power in, 534  
     motor irritation in, 535



- Peripheral nerves diseases of, nails in, 541  
 paresthesia in, 539  
 prognosis of, 541  
 reaction of degeneration in, 535  
 skin in, 540  
 symptoms of, motor, 533  
   sensory, 536  
   trophic, 540  
   vasomotor, 540  
 tension of muscles in, 534  
 treatment of, 541  
 neuritis, 573  
   diagnosis of, from syphilitic myelitis, 242
- Petit mal, 604  
 aura in, 605
- Pharyngeal nerves, paralysis of, 517
- Pharynx in myasthenia gravis, 804
- Phobias, 52
- Phonic spasm, 522
- Photophobia in acute meningitis, 170  
 in trigeminal neuralgia, 734
- Photopsia, 36
- Phrenic nerve, paralysis of, 543
- Pineal gland, lesion of, 291  
 tumors of, 336
- Pithiatism, 659
- Pituitary glands, lesion of, 291
- Pleurisy in arthritis deformans, 915
- Pleurodynia, 945
- Plexiform neuromata, 571
- Pneumogastric nerve, diseases of, 517
- Pneumonia, acute meningitis and, 168
- Poisoning, opium, diagnosis of, from apoplexy, 442  
 tetany and, 749
- Poliencephalitis, acute inferior, 368  
 diagnosis of, from myasthenia gravis, 806  
 hemorrhagic superior, 365  
   alcoholic, 366  
     diagnosis of, 367, 373  
     gait in, 367  
     headache in, 366  
   non-alcoholic, 365  
   ophthalmoplegia in, 365
- Poliencephalomyelitis, 369
- Poliomyelitis, acute, diagnosis of, from hematomyelia, 263  
 from neuritis, 586  
 from progressive spinal muscular atrophy, 124  
 anterior, chronic, 82  
   diagnosis of, from progressive muscular dystrophy, 124  
   neural muscular atrophy, 111  
 diagnosis of, from Landry's paralysis, 252  
 facial paralysis and, 497
- Polyarthritides, multiple neuritis and, 585
- Polychromates, 25
- Polydipsia in tumor of brain, 338
- Polyideic somnambulism, 44
- Polymyositis hemorrhagica, 879
- Polymyositis hemorrhagica, diagnosis of, 880  
 etiology of, 879  
 oedema in, 879  
 pathology of, 879  
 prognosis of, 880  
 skin in, 879  
 symptoms of, 879  
 treatment of, 880
- Polyneuritic psychosis, 45
- Polyneuritis, 573  
 acute infective, 590  
   course of, 590  
   onset of, 590  
   paralysis in, 590  
   spleen in, 590  
   symptoms of, 590  
 facial paralysis and, 497  
 recurring, 591
- Polyuria in brain tumor, 337, 338, 342
- Pons, hemorrhage of, diagnosis of, from progressive bulbar palsy, 117  
 lesion of, 289  
   coma in, 290  
   convulsions in, 290  
   headache in, 290  
   hemianesthesia in, 290  
   hemiplegia in, 290  
   paralysis in, 289  
   vertigo in, 290  
 tumors of, 336  
   diagnosis of, from progressive bulbar palsy, 117
- Pontile syndrome, 289
- Pontomedulloccerebellar space, lesion in, 289
- Popliteal nerves, paralysis of, 559
- Porencephaly, apoplexy and, 409  
 epilepsy and, 598
- Position, anomalies of, 30
- Posterior spinal sclerosis, 833. *See* Tabes dorsalis.
- Posthemiplegic chorea, 466
- Pott's disease, 188. *See* Spinal caries.
- Pregnancy, apoplexy and, 423, 425  
 arthritis deformans and, 902  
 chorea and, 777, 779  
 myasthenia gravis and, 799  
 osteomalacia and, 948  
 progressive spinal muscular atrophy and, 81
- Presbyopia, 36
- Pressure sense of, 19, 32
- Professional cramps, 741  
 neuroses, 741
- Progressive bulbar palsy, 113  
 muscular dystrophy, 117  
   diagnosis of, from progressive muscular atrophy, 86  
 spinal muscular atrophy, 82  
   of childhood, 87
- Propulsion, 74
- Protopathic sensibility, 17
- Psammoma of brain, 318
- Psammosarcoma of brain, 315
- Pseudo-achondroplasia, 969
- Pseudo-bulbar paralysis, 290, 798

- Pseudo-hallucinations, 37  
 Pseudo-hypertrophic progressive muscular dystrophy, 122  
 Pseudo-paralytica, 798  
 Pseudo-paresis, alcoholic, diagnosis of, from paresis, 860  
   syphilitic, diagnosis of, from paresis, 860  
 Pseudo-sclerosis, 158  
   diagnosis of, 159  
   epileptic attacks in, 159  
   etiology of, 158  
   nystagmus in, 159  
   pain in, 159  
   pathology of, 158  
   reflexes in, 159  
   speech in, 159  
   symptoms of, 159  
   tremors in, 159  
   weakness in, 159  
 Pseudo-tabes, hysteria and, 680  
   syphilitica, 826  
 Pseudo-tetany, hysterical, diagnosis of, from tetany, 760  
 Psychic blindness, 299  
   disturbances in brain tumor, 329, 342  
   in migraine, 719  
   epilepsy, 602  
   factors in occupation neuroses, 742  
   influences, epilepsy and, 597  
   manifestations in chorea, 780  
   neuralgia, 726  
   symptoms of Huntington's chorea, 789  
 Psychoses in hysteria, 689  
   post-epileptic, 603  
   traumatic, 642  
     diagnosis of, 648  
     prognosis of, 652  
 Ptosis in cerebrospinal syphilis, 825  
   in hemiplegia, 459  
   in hysteria, 679  
   in ocular paralysis, 488  
   in paralyzing vertigo, 515  
 Pudendal plexus, neuralgia of, 740  
 Pulmonary nerves, paralysis of, 523  
 Pulse in abscess of brain, 380  
   in acute meningitis, 170  
   in apoplexy, 435  
   in arthritis deformans, 909, 914  
   in cervical ribs, 556  
 Pupillary reaction, hemianopic, 482  
   of Wernicke, 271  
   reflexes, anomalies of, 76  
 Pupils in apoplexy, 439  
   in disseminated sclerosis, 152  
   in general paresis, 856  
   in tabes dorsalis, 837  
   variations of, 487  
 Pyorrhœa alveolaris, arthritis deformans and, 901  
 Pyramidal tracts in amyotrophic lateral sclerosis, 90

## Q

- QUADRIGEMINAL bodies, lesion of, 282  
   hearing in, 282

- Quadrigeminal bodies, lesion of, ophthalmoplegia in, 283  
   vision in, 282  
   tumor of, 282

## R

- RADIAL nerve, paralysis of, 547  
 Raynaud's disease, arthritis deformans and, 929  
 Reaction, anomalies of mode of, 48  
   of degeneration, 72  
     in amyotrophic lateral sclerosis, 99  
     distal, 535  
     myasthenic, 73  
     myotonic, 73, 891  
 Recording faculty, examination of, 45  
 Rectal reflex in lesions of spinal cord, 182  
 Rectus muscles, external, paralysis of, 485  
   inferior, paralysis of, 485  
   internal, paralysis of, 485  
   superior, paralysis of, 485  
 Reflex hypothesis of migraine, 710  
   irritation, convulsions and, 792  
   neuralgia, 726  
 Reflexes in amaurotic family idiocy, 869  
   in amyotrophic lateral sclerosis, 97  
   in apoplexy, 439  
   in arthritis deformans, 917  
   in chorea, 781  
   in diseases of peripheral nerves, 534  
   in disseminated sclerosis, 149  
   in general paresis, 857  
   in hemiplegia, 465  
   in hysteria, 688  
   in lesion of cerebellum, 286  
   in multiple neuritis, 584  
   in myasthenia gravis, 805  
   in myotonia congenita, 892  
   in neurasthenia, 632  
   in osteomalacia, 951  
   in paralysis agitans, 771  
   in periodic paralysis, 814  
   in progressive bulbar palsy, 116  
     neural muscular atrophy, 110  
     spinal muscular atrophy, 85  
   in pseudo-sclerosis, 159  
   in spondylitis deformans, 927  
   in tabes dorsalis, 837  
   in tetany, 759  
 Reproduction, anomalies of, 53  
 Resistance, sensation of, 17  
 Respiration in abscess of brain, 380  
   in apoplexy, 436  
   in hemiplegia, 460  
   in Paget's disease, 976  
 Retina, lesions of, 474  
 Retinitis, 474  
   albuminuric, 475  
   syphilitic, 475  
 Retrobulbar neuritis, 37, 479  
 Retropulsion, 74  
 Rhachitis annularis, 978  
 Rheumatic fever, chorea and, 775

Rheumatic fever, diagnosis of, from arthritis deformans, 930  
 Rheumatoid arthritis, 897, 908  
 Rhinogenic abscess of brain, 385  
 Ribs in achondroplasia, 957  
     cervical, 554  
 Rickets, achondroplasia and, 966  
     annular, 955  
     congenital, 955  
     convulsions and, 792, 793  
     diagnosis of, from hydrocephalus, 355  
     fetal, 955  
     intra-uterine, 955  
     micromelic, 955  
 Rigidity in compression of spinal cord, 190  
 Rinne's test in nerve deafness, 510  
 Romberg symptom in tabes dorsalis, 838  
 Rupture of cerebral arteries, 400  
 Rhythmic movements in hysteria, 676

## S

SACRAL cord, disease of, diagnosis of, from paralysis of nerves, 561  
     plexus, diseases of, diagnosis of, from paralysis of nerves, 561  
     paralysis of, 560  
 Sacro-iliac disease, diagnosis of, from sciatica, 565  
     joint in arthritis deformans, 912, 923  
 Sacrum, disease of, diagnosis of, from sciatica, 565  
 Sadness, pathological, 60  
 St. Vitus' dance, 774  
 Salivation in migraine, 718  
 Salvarsan in cerebrospinal syphilis, 830  
     intraspinal, 831, 862  
 Salvarsanized serum, 831, 862  
 Sarcoma of brain, 313, 314, 315  
     of spinal cord, 197  
 Sarcomatosis, diagnosis of, from syphilis, 827  
 Scanning speech, 74  
 Scapular reflex, 78  
 Scapulodynia, 945  
 Scarlet fever, apoplexy and, 417  
     multiple neuritis and, 575  
 Schwabach's test, 24  
 Sciatic nerve, paralysis of, 558, 560  
     in spondylitis deformans, 927  
 Sciatica, 562  
     diagnosis of, 565  
     from disease of cauda equina, 565  
     of sacrum, 565  
     from hip disease, 565  
     from intermittent claudication, 565  
     from pelvic tumors, 565  
     from sacro-iliac disease, 565  
     from tabes dorsalis, 565  
     etiology of, 562  
     fever in, 563  
     Lasègue's sign in, 563  
     organic, diagnosis of, from sciatic neuritis, 565  
     pain in, 562

Sciatica, paresthesia in, 563  
     prognosis of, 565  
     simple, diagnosis of, from sciatic neuralgia, 565  
     symptoms of, 562  
     treatment of, 566  
 Scleroderma, arthritis deformans and, 929  
 Sclerosis, amyotrophic lateral, 89  
     bulbar changes in, 92, 100  
     contractures in, 99  
     diagnosis of, 101  
     from progressive spinal muscular atrophy, 86  
     duration of, 94  
     etiology of, 89  
     frequency of, 90  
     lead and, 89  
     mental failure in, 99  
     muscles in, 100  
     pain in, 99  
     pathology of, 90  
     prognosis of, 101  
     pyramidal tracts in, 90  
     reflexes in, 98  
     reaction of degeneration in, 99  
     symptoms of, 93  
     syphilis and, 90  
     treatment of, 101  
     tremors in, 98  
     types of, 95  
         amyotrophic, 95  
         labio-glossolaryngeal, 95  
         ordinary, 95  
         spastic, 96  
         weakness in, 97  
 of brain, 140  
 of cerebellum, 287  
 combined, diagnosis of, from lateral sclerosis, 104  
     from tabes dorsalis, 845  
 diffuse, 160  
 disseminated, 140  
     age and, 141  
     ataxia in, 147, 154  
     Babinski's sign in, 149  
     cerebral, 154  
     cerebrospinal, 154  
     classical, 154  
     course of, 157  
     defecation in, 150  
     diagnosis of, 154  
     from brain tumor, 343  
     from cerebrospinal syphilis, 156  
     from Friedreich's ataxia, 138  
     from general paresis, 156, 860  
     from hysteria, 155, 692  
     from lateral sclerosis, 104  
     from myelitis, 155  
     from paralysis agitans, 772  
     from spastic paraplegia, 156  
     from subacute combined degeneration, 155  
     from syphilis, 827  
     from tabes dorsalis, 845

- Sclerosis, disseminated, diagnosis of, from
- tumor of brain, 156
  - diplopia in, 150
  - emotion and, 143
  - epileptic attacks in, 153
  - etiology of, 141
  - frequency of, 140
  - gait in, 148
  - heredity and, 142
  - histology of, 144
  - hypertonicity in, 147
  - hysterical, 154
  - infections and, 141
  - intention tremor in, 148
  - intermittent, 154
  - metallic poisons and, 142
  - micturition in, 150
  - nystagmus in, 151
  - ocular, 154
    - muscles in, 151
  - onset of, 154
  - optic, 154
    - disks in, 151
  - pain in, 149
  - pathogenesis of, 145
  - pathology of, 143
  - prognosis of, 156
  - pupil in, 152
  - reflexes in, 149
  - sacral, 154
  - speech in, 152
  - spinal, 154
  - symptoms of, 147
    - mental, 153
    - motor, 147
    - sensory, 148
  - syphilis and, 142
  - trauma and, 142
  - treatment of, 157
  - trigeminal neuralgia in, 733
  - trophic changes in, 153
  - types of, 154
  - vasomotor changes in, 153
  - vision in, 150
  - weakness in, 147, 154
- insular, 140
- lateral, 101
  - diagnosis of, 104
    - from caries of spine, 104
    - from combined sclerosis, 104
    - from hysteria, 104
    - from multiple sclerosis, 104
    - from myelitis, 104
    - from secondary degeneration, 104
    - from tumors of spinal cord, 104
  - pathology of, 101
  - spasticity in, 102
  - symptoms of, 102
  - treatment of, 104
  - types of, 103
  - weakness in, 102
- miliary, 161
- multiple, 140
- posterior spinal, 833. *See* Tabes dorsalis.
- Sclerosis, subacute combined, of cord, 126
  - anemia and, 128
  - ataxic paraplegia in, 127
  - diagnosis of, 129
    - from tabes, 129
  - duration of, 129
  - etiology of, 126
  - fever in, 129
  - flaccid paraplegia in, 128
  - onset of, 128
  - pathology of, 126
  - prognosis of, 130
  - spastic paraplegia in, 128
  - spasticity in, 127
  - symptoms of, 127
  - treatment of, 130
- tuberous, 160
- Scotoma, 34
- Scotomata in migraine, 712, 714
- Secretions in migraine, 718
- Secretory disturbances in hysteria, 687
  - in neuralgia, 728
  - in syringomyelia, 224
- functions, anomalies of, 79
- Senile dementia, trauma and, 647
- Sensation, deep, 536
  - disturbances of, 28
  - epicritic, 537
  - protopathic, 537
- Sense of acceleration, 18
  - of equilibrium, 18
  - exterior, 19
  - of fatigue, 19, 32
  - of hearing, 22
  - of hunger, 19, 31
  - interior, 17
  - of itching, 20
  - of movement, 30
  - muscle, 17
  - pain, 17
    - anomalies of, 29
  - perception, 18, 30
  - of position, anomalies of, 30
  - of pressure, 19, 32
  - of resistance, 17, 30
  - sexual, 19, 32
  - of sight, 24
  - of smell, 21, 33
  - of taste, 21, 33
  - temperature, 20, 32
  - of thirst, 19, 32
  - of tickling, 20
  - of touch, 19, 32
  - vestibular, 18, 30
  - vibration, 18, 30
- Sensibility, deep, 17, 184
  - anomalies of, 30
  - disturbances of, in lesions of the spinal cord, 183
  - epicritic, 17, 184
  - mesoblastic, 17
    - anomalies of, 30
  - muscular, in tabes dorsalis, 839
  - osseous, 30
  - protopathic, 17, 184
  - stereognostic, 18, 30

- Sensory aphasia, 74, 292, 296
  - area of cortex of brain, 265
  - disturbances in occupation neuroses, 747
  - phenomena in tetany, 759
- Sexual reflex in spinal cord lesions, 182
  - sense, 19, 32
- Shaking palsy, 764
- Shoulder-joint in arthritis deformans, 911
- Sick headache, 709
- Sight, anomalies of, 34
  - sense of, 24
- Simulation, in hysteria, 659, 667
- Sinus thrombosis, brain abscess and, 378
  - diagnosis of, from brain abscess, 386
  - from encephalitis, 374
- Skin in arthritis deformans, 916
  - in diseases of peripheral nerves, 540
  - in facial hemiatrophy, 992
  - in herpes zoster, 572
  - in hysteria, 686
  - in neuralgia, 728
  - in polymyositis hemorrhagica, 879
  - in syringomyelia, 224
  - in trigeminal neuralgia, 734
- Skull in achondroplasia, 956
  - hyperostosis of, 988
  - in Paget's disease, 971
- Smallpox, multiple neuritis and, 575
- Smell, anomalies of, 33
  - centre of, 271
  - disturbances of, 471
    - diagnosis of, 472
    - etiology of, 471
    - pathology of, 472
    - prognosis of, 473
    - symptoms of, 472
  - in hysteria, 684
  - sense of, 21
    - anomalies of, 33
- Softening, central, onset of, 431
- Somatopsyche, 42
- Somnambulisms, monoideic, 44
  - polyideic, 44
- "Soul paralysis," 268
- Spasm, habit, 784
  - of muscles of neck, 525
  - of tongue, 532
  - phonic, 522
- Spasmodic torticollis, 526
- Spasmophilia, 604
- Spastic conjugate deviation in apoplexy, 438
  - dysphonia, 522
  - spinal paralysis, 101
    - hereditary, 105
  - syringomyelia, 220
  - tabes, 101
- Spastic-paretic gait, 73
- Speech, anomalies of, 74
  - in disseminated sclerosis, 152
  - in epilepsy, 612
  - in general paresis, 856
  - in lesion of centrum semiovale, 275
  - in migraine, 715
  - in progressive bulbar palsy, 115
- Speech in pseudo-sclerosis, 159
- Sphincters in multiple neuritis, 584
- Spina bifida, hydrocephalus and, 354
- Spinal accessory nerve, paralysis of, 524
  - ataxic gait, 73
  - caries, 188
    - diagnosis of, from lateral sclerosis, 104
    - from tumors, 208
  - dry, 189
  - etiology of, 188
  - pathology of, 188
- cord, abscess of, 254
  - anemia of, 255
  - carcinoma of, 196
    - diagnosis of, from caries, 193
  - cavities of, 210
  - combined system, diseases of, 125
  - compression of, 188
    - from caries, anesthesia in, 191
    - course of, 192
    - deformity in, 190
    - diagnosis of, 192
      - from congenital abnormalities, 193
      - from functional paraplegia, 194
      - from syphilitic meningomyelitis, 193
      - from syringomyelia, 193
    - extension of, 195
    - fever in, 192
    - onset of, 192
    - pain in, 190
    - prognosis in, 194
    - prophylaxis in, 194
    - rest in, 195
    - rigidity in, 190
    - root pains in, 191
    - symptoms of, 190
    - treatment of, 194
      - operative, 195
    - weakness in, 191
- congenital abnormalities of, diagnosis of, from caries, 193
- cysts of, diagnosis of, from tumors, 208
- diseases of, 178
  - focal, differentiation of, 187
- embolism in, 256
- fissures of, 210
- hemorrhage into, 260
- hyperemia of, 257
- impulses in, 184
- lesions of, 178, 180
  - Brown-Séquard paralysis in, 185
  - extramedullary, 187
  - flaccid paralysis in, 178
  - intramedullary, 187
  - oculopupillary symptoms and, 181
  - optic neuritis in, 181
  - pain in, 183
  - paralysis in, 178, 179
  - reflexes in, 182

- Spinal cord, lesions of, secretory phenomena in, 181  
 sensibility in, 183  
 spastic paralysis in, 179  
 symptoms of, cardiac, 182  
     respiratory, 182  
     splanchnic, 182  
 vasomotor phenomena in, 181  
 myeloma of, 198  
 sacral, diseases of, diagnosis of, from paralysis of nerves, 561  
 sarcoma of, 197  
 sclerosis of, combined, caused by vegetable poisons, 139  
 segment of, as centre of activity, 180  
 segmental localization of, 178  
 spinal root symptoms of, 180  
 syphilis of, 823. *See* Syphilis, cerebrospinal.  
 thrombosis in, 256  
 tumors of, 196  
     Brown-Séquard paralysis in, 204  
     cauda equina, 206  
     cervical enlargement, 204  
     course of, 206  
     diagnosis of, 207  
     from caries, 208  
     from cervical pachymeningitis, 208  
     from cysts, 208  
     from lateral sclerosis, 104  
     from tabes dorsalis, 845  
 dorsal, 205  
 intravertebral, 198  
     extradural, 198  
     incidence of, 200  
     intradural, 198  
     intramedullary, 199  
     pathology of, 199  
 lumbosacral, 205  
 pain in, 201, 202  
 paraplegia in, 203  
 prognosis of, 209  
 result of, 206  
 site of, 201  
 symptoms of, 201  
     sensory, 203  
     spinal cord, 203  
     root, 202  
     vertebral, 201  
 treatment of, 209  
 of upper cervical region, 204  
 vertebral, 196  
     benign, 198  
     vascular disorders of, 255  
 curvature in syringomyelia, 222  
 disseminated sclerosis, 154  
 membranes, hemorrhage into, 257  
 meningitis, localized serous, 174  
 muscular atrophy, progressive, 82, 87  
 nerves, diseases of, 542  
 paralysis, spastic, 101  
     hereditary, 105
- Spinal paralysis, syphilitic, 828  
 roots, diseases of, 567  
     diagnosis of, 568  
     onset of, 567  
     symptoms of, 567  
     treatment of, 568  
 sclerosis, posterior, 833. *See* Tabes dorsalis.  
 Spine in Paget's disease, 971  
 Spirochæta pallida in general paresis, 851, 852  
     in nervous system, 823  
     in tabes dorsalis, 835  
 Spleen in acute infective polyneuritis, 590  
     in arthritis deformans, 909, 915  
 Spondylitis deformans, 923  
     age and, 925  
     atrophy in, 927  
     diagnosis of, 928  
     from Paget's disease, 977  
     etiology of, 925  
     occurrence of, 925  
     pain in, 925, 926  
     reflexes in, 927  
     sciatic nerve in, 927  
     signs of, 926  
     symptoms of, 924  
     tendo-Achilles reflex in, 927  
     trauma and, 925  
     x-rays in, 927  
 Stammering, 74  
 Staphylococcus pyogenes aureus in pyogenic encephalitis, 363  
 Status epilepticus, 609  
     hemicranus in migraine, 718  
 Stereognosis, anomalies of, 30  
 Stereognostic sense, 18  
 Sternoclavicular joint in arthritis deformans, 911  
 Still's disease, 917, 931  
 Strabismus in ophthalmoplegia, 484  
 Stupor, 47, 63, 65  
 Stuttering, 74  
 Subacute combined sclerosis of spinal cord, 126  
 Subcutaneous nodules in arthritis deformans, 915  
 Sunstroke, diagnosis of, from apoplexy, 444  
 Superior pontile syndrome, 291  
 Suprascapular nerve, paralysis of, 545  
 Sydenham's chorea, 774  
 Syndrome, Benedikt's, 369  
     cerebellar deficit, 287  
     Millard-Gubler, 369  
     pontile, 289  
     superior, 291  
     tegmental, 281  
     thalamic, 279, 334, 467  
     Weber's, 369  
 Synesthesia, 29  
 Synovial membrane in arthritis deformans, 904, 905  
 Syphilis, achondroplasia and, 967  
     amyotrophic lateral sclerosis and, 90  
     apoplexy and, 423, 425, 426, 427  
     arteriosclerosis of brain and, 393

- Syphilis of brain, 823. *See* Syphilis, cerebrospinal.
- cerebral aneurism and, 399
- thrombosis and, 400, 401
- cerebrospinal, 823, 824
- deafness in, 825
- diagnosis of, 827
- from carcinomatosis, 827
- from disseminated sclerosis, 156, 827
- from sarcomatosis, 827
- from tabes, 826
- from tuberculosis, 827
- endarteritis in, 824
- etiology of, 824
- headache in, 825
- meninges in, 824
- ophthalmoplegia in, 825
- paralysis in, 825
- paraplegia in, 825
- ptosis in, 825
- salvarsan in, 830
- symptoms of, 825
- treatment of, 830
- intraspinal, 831
- vomiting in, 825
- Wassermann reaction in, 826, 827
- chorea and, 775, 777
- epilepsy and, 595
- general paresis and, 850
- hereditary, hemiplegia in, 418
- myelitis and, 228
- of nervous system, 822
- neuralgia and, 727
- neurasthenia and, 629
- in ocular paralysis, 488
- osteitis deformans and, 973
- osteo-arthropathy and, 982
- postnatal infantile hemiplegia and, 418
- of spinal cord, 823. *See* Syphilis, cerebrospinal.
- syringomyelia and, 212
- tabes dorsalis and, 833
- Syphilitic choroidoretinitis, 475
- meningomyelitis, diagnosis of, from
- tabes dorsalis, 845
- myelitis, 237, 828
- paraplegia, Erb's, 246, 828
- pseudoparesis, diagnosis of, from
- paresis, 860
- refinitis, 475
- spinal paralysis, 828
- Syphiloma of brain, 320, 321
- Syringomyelia, 211
- age and, 211
- amyotrophic, 220
- analgesia in, 223
- anesthesia in, 223
- bones in, 225
- bulbar symptoms in, 222
- cavity in, 212
- central canal in, 214
- congenital anomalies and, 211
- definition of, 211
- diagnosis of, 226
- from caries of spinal cord, 193
- from hematomyelia, 263
- Syringomyelia, diagnosis of, from hysteria, 693
- from progressive muscular atrophy, 226
- from spinal muscular atrophy, 86
- from tabes dorsalis, 845
- etiology of, 211
- gliomatosis in, 213
- heredity and, 211
- incoördination in, 221
- joints in, 225
- Morvan's type, 220
- nervous elements in, 214
- nystagmus in, 222
- onset of, 220
- paralysis in, atrophic, 221
- ocular sympathetic, 224
- spastic, 221
- pathogenesis of, 215
- pathology of, 212
- prognosis of, 227
- secretory disturbances in, 224
- sensation in, 222
- skin in, 224
- spastic, 220
- sphincters in, 226
- spinal curvature in, 222
- symptoms of, 218
- syphilis and, 212
- thermo-anesthesia in, 222
- trauma and, 211
- treatment of, 226
- tremors in, 222
- trophic disturbances in, 224
- urticaria in, 224
- vascular lesions in, 214
- vasomotor disturbances in, 224

## T

## TABES amaurotica, 844

atactica, 844

dolorosa, 844

dorsalis, 833

Achilles tendon reflex in, 837

age and, 834

alcoholia in, 838

anesthesia in, 838

Argyll-Robertson pupil in, 836

arthropathy in, 842

astereognosis in, 838

ataxia in, 839

atrophy of optic nerve in, 841

bladder in, 840

cardiac crises in, 842

course of, 843

cranial nerves in, 841

definition of, 833

diagnosis of, 844

from brain tumor, 845

from cerebrospinal syphilis, 826

from combined sclerosis, 845

from Friedreich's ataxia, 138, 845

from multiple neuritis, 845

- Tabes dorsalis, diagnosis of, from multiple sclerosis, 845  
     from neuralgia, 730  
     from neuritis, 587  
     from sciatica, 565  
     from spinal tumor, 845  
     from subacute combined sclerosis, 129  
     from syphilitic meningo-myelitis, 845  
     from syringomyelia, 845  
     duration of, 843  
     etiology of, 833  
     facial nerves in, 841  
     gastric crises in, 841, 849  
     hypalgesia in, 838  
     hyperesthesia in, 836, 838  
     intestinal crises in, 842  
     knee-jerks in, 837  
     migraine and, 720  
     muscular hypotonia in, 839  
         sensitivity in, 839  
     mydriasis in, 837  
     neuralgia and, 727  
     neurasthenia in, 843  
     oculomotor palsies in, 841  
     ophthalmoplegia in, 841  
     pain in, 836  
     paresis and, 843  
     pathology of, 834  
     prognosis of, 846  
     prophylaxis in, 847  
     pupils in, 837  
     Romberg symptom in, 838  
     sensation in, 838  
     Spirochæta pallida in, 835  
     superficial reflexes in, 837  
     symptoms of, 835  
     syphilis and, 833  
     treatment of, 847  
     trigeminal nerve in, 841  
         neuralgia in, 733  
     trophic disorders in, 841  
     visceral crises in, 841  
     Wassermann reaction in, 847  
     incompleta, 844  
     juvenilis, 844  
     paralytica, 844  
     paresthetica, 844  
     spastic, 101  
     superior, 840, 844  
     visceralis, 844  
 Taste in hysteria, 684  
     sense of, 21  
 Tay-Sachs disease, 863  
 Tegmental syndrome, 281  
 Tegmentum, lesion of, 281  
     paralysis in, 281  
     syndrome of, 281  
     tumors of, 336  
 Temporal lobe, tumors of, 331  
 Temporomaxillary joint in arthritis de-formans, 911  
 Tendo-Achilles reflex in spondylitis de-formans, 927  
 Teratoma of brain, 316  
 Test, association, 48  
 Testicular neuralgia, 740  
 Tetany, 748  
     benign, 759  
     calcium and, 751  
     Chvostek's sign in, 758  
     course of, 759  
     definition of, 748  
     diagnosis of, 760  
         from hysterical pseudotetany, 760  
     electrical hyperexcitability in, 757  
     epidemic, 750, 762  
     Erb's sign in, 757  
     etiology of, 750  
     gastric disease and, 749  
     grave, 760  
     hysteria and, 677  
     idiopathic, 749  
     incidence of, 750  
     infectious diseases and, 749  
     intestinal disease and, 749  
     laryngospasm in, 756  
     maternity and, 749  
     mechanical irritability in, 758  
     mental phenomena in, 758  
     motor nerves in, 757  
     muscle cramps in, 754  
     nervous disease and, 749  
     osteomalacia and, 949  
     pain in, 757, 759  
     parathyreopriva, 761  
     parathyroids and, 749, 750, 753  
     pathology of, 753  
     poisoning and, 749  
     prognosis of, 761  
     reflexes in, 759  
     sensory phenomena in, 759  
     severe, 760  
     sight in, 759  
     strumipriva, 761  
     symptoms of, 754  
     treatment of, 762  
     trophic phenomena in, 759  
     Trousseau's phenomena in, 756  
 Thalamic syndrome, 279, 334, 467  
 Thermanesthesias, 32  
 Thermo-anesthesia in syringomyelia, 222  
 Thirst, sense of, 19, 32  
 Thomsen's disease, 886  
 Thoracic nerve, long, paralysis of, 544  
 Thought inhibition, 47  
 Thrombosis, apoplexy and, 423, 425  
     of cerebral arteries, 400  
         diagnosis of, from tumor, 343  
     in cervical ribs, 556  
     of spinal cord, 256  
 Thumb in arthritis deformans, 922  
 Thymus gland, convulsions and, 794  
     in myasthenia gravis, 799  
 Thyroid gland, achondroplasia and, 965  
     in osteomalacia, 948  
 Tibial nerve, posterior, paralysis of, 559  
 Tic convulsif, 504, 784  
     etiology of, 504  
     treatment of, 505  
         douloureux, 493  
 Tinnitus aurium, 508  
     diagnosis of, 509



- Tinnitus aurium, treatment of, 509  
in vestibular disease, 511
- Toes in arthritis deformans, 913
- Tone deafness in aphasia, 297
- Tongue, atrophy of, 531  
in hemiplegia, 460  
muscles of, spasm of, 532  
paralysis of, 530  
in hysteria, 679
- Tonsillitis, arthritis deformans and, 901
- Tonus of muscles, 68
- Topalgia in hysteria, 683
- Topical diagnosis of diseases of brain, 265
- Torticollis, 526, 945  
spasmodic, 526  
etiology of, 526  
pain in, 528  
pathology of, 528  
prognosis of, 528  
symptoms of, 528  
treatment of, 529
- Touch, sense of, 19, 32
- Toxemia, apoplexy and, 425
- Toxic hypothesis of migraine, 710  
myelitis, 236
- Toxins, epilepsy and, 594  
myasthenia gravis and, 800  
neuralgia and, 726
- Trauma, acute mania and, 647  
apoplexy and, 424  
arthritis deformans and, 901  
diagnosis of, from apoplexy, 444  
disseminated sclerosis and, 142  
epileptic insanity and, 647  
general paresis and, 647  
hysteria and, 657  
hysterical insanity and, 647  
paralysis agitans and, 764  
paranoia and, 647  
senile dementia and, 647  
spondylitis deformans and, 925  
syringomyelia and, 211  
villous arthritis and, 929
- Traumatic abscess of brain, 382  
hysteria, 644  
lumbago, 644  
mental disturbances, 646  
neurasthenia, 643  
neuroses, 642  
psychoses, 642
- Tremor, 70  
in amyotrophic lateral sclerosis, 98  
in brain tumor, 340  
in general paresis, 856  
in hemiplegia, 465  
in hysteria, 676  
in paralysis agitans, 768  
in progressive spinal muscular atrophy, 85  
senile, diagnosis of, from paralysis agitans, 773  
in syringomyelia, 222
- Trichiniasis, diagnosis of, from neuritis, 587
- Trigeminal nerve, anatomy of, 491  
diseases of, 491  
pathology of, 492  
symptoms of, 493
- Trigeminal nerve, diseases of, taste and, 493  
tic douloureux in, 493  
maxillary branches, 491  
motor paralysis of, 494  
ophthalmic branches, 491  
paralysis of, 493, 494  
in tabes dorsalis, 841  
trismus and, 495  
neuralgia, 733
- Trismus, 495
- Trochlear nerve, anatomy of, 483
- Trophic changes in disseminated sclerosis, 153  
in general paresis, 858  
in hysteria, 685  
in migraine, 718  
in multiple neuritis, 584  
in syringomyelia, 224  
in tabes dorsalis, 841  
functions, anomalies of, 79  
phenomena in tetany, 759
- Trousseau's phenomenon, 71  
in tetany, 756
- Tubercle of brain, solitary, 319
- Tubercula dolorosa, 570
- Tuberculoma of brain, encephalic, 319
- Tuberculosis, cerebrospinal, diagnosis of, from syphilis, 827  
multiple neuritis and, 575, 586  
osteo-arthropathy and, 982
- Tuberous sclerosis, 160
- Tumors of basal ganglia, 334  
of brain, 308  
stem, 335  
of cavum Meckelii, 340  
of centrum semiovale, 276, 334  
of cerebellum, 338  
of corpora quadrigemina, 336  
of corpus callosum, 276, 335  
of cranial base, 340  
of crura, 335  
extracerebellar, 338  
of fourth ventricle, 338  
of frontal lobe, 331  
hydrocephalus and, 356  
intracerebellar, 338  
intravertebral, of spinal cord, 198  
of medulla oblongata, 337  
of nerves, 568  
Paget's disease and, 973  
of paracentral convolution, 332  
of parietal lobe, 333  
of pineal gland, 336  
of pons, 336  
of quadrigeminal bodies, 282  
of spinal cord, 196  
of tegmentum, 336  
of temporal lobe, 331  
vertebral, of spinal cord, 196
- Twilight states, 44
- Twitchings, fibrillary, 70
- Typhoid fever, apoplexy and, 417, 425  
cerebral thrombosis and, 400  
disseminated sclerosis and, 141  
multiple neuritis and, 575  
neurasthenia and, 629

## U

- ULNAR nerve, paralysis of, 549  
 Uncinate fits, 332  
 Unconsciousness, 43  
 Unilateral paralysis, 108  
 Uremia, diagnosis of, from apoplexy, 441  
 Urine in arthritis deformans, 916  
   in migraine, 718  
   in myasthenia gravis, 805  
   in neurasthenia, 632  
   in osteomalacia, 952  
   in periodic paralysis, 811  
 Urticaria in syringomyelia, 224  
 Uvular reflex, 78

## V

- VAGAL attacks, 523  
 Vagus nerve, branches of, cardiac, par-  
   alysis of, 522  
   gastric, paralysis of, 523  
   laryngeal, paralysis of, 518  
   oesophageal, paralysis of, 523  
   pharyngeal, paralysis of, 517  
   pulmonary, paralysis of, 523  
   diseases of, 517  
   paralysis of, 517  
   vagal attacks and, 523  
 Valleix's points, 17, 727, 734, 737  
 Vascular changes in infective myelitis, 232  
   disorders of spinal cord, 255  
   lesions, diagnosis of, from brain  
   tumor, 343  
 Vasomotor changes in disseminated scler-  
   osis, 153  
   in multiple neuritis, 584  
   disturbances in epilepsy, 606  
   in general paresis, 858  
   in hysteria, 685  
   in Landry's paralysis, 251  
   in migraine, 718  
   in neuralgia, 728  
   in syringomyelia, 224  
   effects in hemiplegia, 468  
   functions, anomalies of, 79  
   hypothesis of hysteria, 662  
   of migraine, 710  
   paresis in Freidreich's ataxia, 136  
 Vertebrae in achondroplasia, 957  
 Vertebral arteries, anatomy of, 387  
 Vertigo, 30, 511  
   in abscess of brain, 379, 384  
   in anemia of brain, 392  
   in arteriosclerosis of brain, 396  
   auditory, 512  
   aural, 513  
   in brain tumor, 329, 338  
   in lesion of pons, 290  
   in neurasthenia, 631  
   paralyzing, 515  
 Vesical reflex in lesions of the spinal  
   cord, 182  
 Vestibular apparatus in hysteria, 684  
   nerve, diseases of, 511  
   nystagmus in, 511  
   tinnitus in, 511  
 Vibration sense, 18

- Villous arthritis, 929  
 Vision, 480  
   acuity of, determination of, 26  
   capacity of, determination of, 27  
   central, 25  
   excentric, 25  
   in hysteria, 652  
   in lesion of centrum semiovale, 275  
   of quadrigeminal bodies, 282  
   in migraine, 712, 714  
   in neurasthenia, 632  
   paracentral, 25  
   in paralyzing vertigo, 515  
   in tetany, 759  
   in traumatic neuroses, 652  
   in trigeminal neuralgia, 734  
 Visual aphasia, 298  
 Voice in paralysis agitans, 771  
 Vomiting in abscess of brain, 379  
   in acute bulbar myelitis, 368  
   meningitis, 170  
   in brain tumor, 326, 338  
   in cerebral aneurism, 399  
   in cerebrospinal syphilis, 825  
   in disease of vagus nerve, 517  
   in Ménière's disease, 513  
   in migraine, 712  
   in pachymeningitis interna, 165  
   in recurrent oculomotor palsy, 488  
 von Graefe's sign in myasthenia gravis, 803  
 von Recklinghausen's disease, 571

## W

- WASSERMANN reaction in cerebrospinal  
   syphilis, 826, 827  
   in general paresis, 851  
   in tabes dorsalis, 847  
 Weakness in lateral sclerosis, 102  
   in progressive muscular dystrophy,  
   120  
   spinal muscular atrophy, 84  
 Weber's syndrome, 369  
   test, 24  
   in nerve deafness, 510  
 Werdnig-Hoffmann type of spinal mus-  
   cular atrophy, 87  
 Wernicke's aphasia, 299  
   disease, 365  
   zone of, 300  
 Whooping cough, apoplexy and, 417  
 Word amnesia, 76  
   blindness, 54, 76, 298  
   in abscess of brain, 382  
   in aphasia, 298  
   deafness, 54, 75, 297  
   in aphasia, 297  
 Wrist-joint in arthritis deformans, 911  
 Writer's cramp, 744  
   paralytic, 746  
   tremulous, 746  
 Writing, anomalies of, 74  
 Wryneck, 526

## X

- X-RAYS in spondylitis deformans, 927

# GENERAL INDEX.

## A

- ABDOMEN** in acute enteritis, iii, 330  
     peritonitis, iii, 678, 680  
     in arteriosclerosis, iv, 464  
     in ascites, iii, 662  
     in chronic enteritis, iii, 335  
     perihepatitis, iii, 491  
     peritonitis, iii, 719  
     in cirrhosis of liver, iii, 522  
     of stomach, iii, 277  
     in mesenteric embolism, iii, 436  
     thrombosis, iii, 436  
     in mucous colitis, iii, 439  
     in rickets, ii, 790  
     in splanchnoptosis, iii, 753  
     in subphrenic pyopneumothorax, iii, 712  
     in tuberculosis of mesenteric glands, iv, 744  
     of peritoneum, iii, 724  
**Abdominal actinomycosis**, i, 1039, 1043  
     heart, iv, 346  
     mediastinal contents, ii, 1109  
     pain in amoebic dysentery, ii, 34, 37  
     in lobar pneumonia, i, 222  
     in typhoid fever, i, 89, 106  
     reflexes, anomalies of, v, 78  
**Aberrant goitre**, iv, 841  
**Abortive plague**, i, 632  
     pneumonia, i, 237  
     smallpox, i, 803  
     typhoid fever, i, 152  
**Abscess in bacillary dysentery**, i, 777  
     of brain, v, 378  
     in amoebic dysentery, ii, 42  
     in bronchiectasis, ii, 917  
     in pleuritis, ii, 1045  
     cervical lymphadenitis and, iv, 733  
     of Fallopian tubes, subphrenic abscess and, iii, 701  
     filarial, ii, 308  
     hepatopulmonary, iii, 503  
     ischio-rectal, in tuberculosis, i, 432  
     of kidney, iii, 937  
     in bronchiectasis, ii, 917  
     of liver, iii, 496  
     in amoebic dysentery, ii, 38  
     in bronchiectasis, ii, 917  
     in typhoid fever, i, 118  
     in lobar pneumonia, i, 217  
     of lung, ii, 993  
     in amoebic dysentery, ii, 43  
     in bronchopneumonia, ii, 914  
     in influenza, i, 538, 540  
     Abscess of lung in lobar pneumonia, i, 250  
         in tuberculosis, i, 430  
     of mesentery, iii, 696  
     metastatic, in gangrene of lung, ii, 991  
     of omentum, iii, 696  
     of pancreas, iii, 635  
     paranephric, iii, 970  
     peri-appendicular, iii, 364, 370  
     pericolic, iii, 697  
     perigastric, iii, 696  
     perinephric, iii, 970  
     peritoneal, residual, iii, 698  
     peritonsillar, i, 705; ii, 849  
     retroperitoneal, iii, 698  
     retropharyngeal, i, 706; ii, 846; iv, 734  
     of spinal cord, v, 254  
     of spleen in amoebic dysentery, ii, 43  
         subphrenic abscess and, iii, 700  
     subphrenic, iii, 699  
         in lobar pneumonia, i, 253  
     thoracic, in tuberculosis, i, 435  
     in typhoid fever, i, 101, 148  
**Absence of adrenal glands**, iv, 781  
     of ductus arteriosus Botalli, iv, 420  
     of pericardium, iv, 77, 344  
     of right superior vena cava, iv, 440  
     of spleen, iv, 938, 944  
     of thoracic duct, iv, 580  
     of thymus, iv, 908  
**Absinthism**, ii, 425  
**Acanthocephali**, ii, 296  
**Acardia**, iv, 350  
**Acariasis**, ii, 317  
**Accessory coronary arteries**, iv, 439  
     pulmonary arteries, iv, 439  
     spleen, iv, 938  
     thymus, iv, 908  
**Acetone bodies in urine**, iii, 826  
     in diabetes mellitus, ii, 702  
**Achilles tendon reflex**, v, 77  
     in tabes dorsalis, v, 837  
**Acholic stools**, iii, 294  
**Achondroplasia**, v, 955  
**Achromates**, v, 25  
**Achromatopsia**, v, 271  
**Achroöcytosis**, iii, 73  
**Achylia in chronic gastritis**, iii, 242  
     gastrica, iii, 29, 153  
     in pernicious anemia, iv, 634  
**Acidity, gastric**, in cholelithiasis, iii, 576  
     in duodenal ulcer, iii, 192, 346  
     in gastric ulcer, iii, 181  
**Acidosis**, ii, 526  
     in hepatic disease, ii, 665

- Acne in congenital syphilis, ii, 190  
 of mouth, iii, 54
- Acoria, iii, 156
- Acrocyanosis, iv, 985
- Acromegaly, iv, 805  
 in exophthalmic goitre, iv, 869  
 glycosuria and, ii, 677, 684
- Acroparesthesia, v, 32
- Acroteric scleroderma, iv, 1024
- Actinomyces bovis, i, 1033
- Actinomyces kidney infections and, iii, 952
- Actinomycoma of brain, v, 321
- Actinomycosis, i, 1032  
 abdominal, i, 1039, 1043  
 of appendix, iii, 386  
 cutaneous, i, 1040  
 of head, i, 1038, 1043  
 of liver, iii, 541  
 of mouth, iii, 59  
 of myocardium, iv, 145  
 of neck, i, 1038, 1043  
 of oesophagus, iii, 86  
 of pleura, ii, 1050  
 thoracic, i, 1039, 1043  
 of thyroid gland, iv, 830
- Acts, anomalies of, v, 66
- Adams-Stokes' syndrome, iv, 80
- Addison's disease, i, 370; iv, 786  
 in tuberculosis, i, 436
- Adductor laryngeal paralysis, v, 521
- Adenitis, acute cervical, i, 1012  
 in diphtheria, i, 701  
 in scarlet fever, i, 874  
 in carcinoma of stomach, iii, 208  
 febrile, i, 636  
 gonococcal, i, 761  
 in lobar pneumonia, i, 233  
 in syphilis, ii, 158
- Adenocarcinoma of thyroid gland, iv, 832, 834
- Adenoids, ii, 840  
 diphtheria and, i, 691  
 thymus and, iv, 913, 916, 924
- Adenolipomatosis, symmetrical, v, 818
- Adenoma of intestines, iii, 433  
 of kidney, iii, 1003  
 of lung, ii, 997  
 of pancreas, iii, 649
- Adherent pericarditis, iv, 44, 51, 58
- Adiadokinesis, v, 72  
 in brain tumor, v, 339
- Adiposis cerebri, v, 818  
 dolorosa, v, 817  
 tuberosa simplex, v, 818
- Adiposity, ii, 761
- Adrenal glands, absence of, iv, 781  
 in acromegaly, iv, 808  
 Addison's disease and, iv, 787  
 in alcoholism, ii, 404  
 anemia of, iv, 782  
 anomalies of, iv, 781  
 atrophy of, iv, 788  
 carcinoma of, iv, 784  
 chromaffin tissue of, iv, 777  
 circulatory disturbances of, iv, 782  
 development of, iv, 777
- Adrenal glands, diabetes mellitus and, ii, 682  
 extract of, iv, 778  
 function of, disordered, iv, 779  
 hemorrhage of, iv, 782  
 hyperemia of, iv, 782  
 hypernephroma of, iv, 784  
 hyperplasia of, iv, 781  
 hypertrophy of, iv, 781  
 in hypopituitarism, iv, 817  
 hypoplasia of, iv, 781  
 infections of, iv, 781  
 inflammation of, iv, 783  
 intoxications of, iv, 781  
 sarcoma of, iv, 784  
 softening of, iv, 783  
 syphilis of, iv, 786  
 tuberculosis of, i, 369; iv, 786  
 tumors of, iv, 784
- insufficiency, iv, 779, 793
- Afebrile endocarditis, iv, 163  
 typhoid fever, i, 153
- Affective states, anomalies of, v, 59
- Afferent nerves, respiration and, ii, 813
- Agamodistomum ophthalmobium, ii, 245
- Agamofilaria georgiana, ii, 315  
 oculi, ii, 314
- Agamomeris restiformis, ii, 316
- Agensis of cerebellum, v, 287
- Agglutination, i, 59  
 in thrombosis, iv, 527
- Agglutinins, i, 59  
 in tuberculosis, i, 313  
 in typhoid fever, i, 162
- Agitation, affective, v, 65  
 hallucinatory, v, 65  
 motor, v, 63
- Aglossia, iii, 60
- Agnosia, v, 269
- Agraphia, v, 75  
 in aphasia, v, 297, 298  
 in migraine, v, 715
- Agusia, iii, 43
- Air embolism, iv, 559  
 passages, upper, obstruction of, ii, 816  
 pathological effects of, ii, 343
- Akinesia algera, v, 816
- Akro-elephantiasis ossea, v, 982
- Albumin in pleural fluid, ii, 1014
- Albuminuria, iii, 794  
 in acute infective polyneuritis, v, 590  
 nephritis, iii, 862  
 in angioneurotic edema, iv, 1008  
 in appendicitis, iii, 371  
 in arteriosclerosis, iv, 463  
 in ascites, iii, 664  
 in chlorosis, iv, 651  
 in chronic nephritis, iii, 882, 902  
 circulatory disturbances and, iii, 795  
 in cirrhosis of liver, iii, 526  
 in congenital cyanosis, iv, 337  
 in diabetes mellitus, ii, 703  
 in diphtheria, i, 701  
 febrile, iii, 795, 867  
 functional, iii, 795  
 gastro-intestinal, ii, 513  
 gonococcal, i, 762

- Albuminuria in gout, ii, 748  
   in hay fever, ii, 864  
   in infantile convulsions, v, 795  
   in infectious jaundice, i, 1016  
   jaundice and, iii, 796  
   kidney and, iii, 774  
     of pregnancy and, iii, 868  
   in lead poisoning, ii, 365  
   in leukemia, iv, 666, 672  
   in lobar pneumonia, i, 233, 258  
   in mercury poisoning, ii, 380  
   in mitral insufficiency, iv, 236  
   in morphinism, ii, 440  
   in nephropotosis, iii, 756  
   in purpura rheumatica, iv, 703  
   in pyelonephritis, iii, 958  
   in Raynaud's disease, iv, 989  
   in renal infarction, iv, 564  
   in rheumatic fever, i, 998  
   in smallpox, i, 807  
   in syphilis of kidneys, ii, 180  
   in tetanus, i, 578  
   in tuberculosis, i, 390, 434  
     of kidney, iii, 982  
   in typhus fever, i, 930  
 Albuminuric bronchitis, ii, 904  
   retinitis, iii, 911; v, 475  
 Albumoses in carcinoma of stomach, iii, 207  
 Albumosuria, iii, 820  
 Alcohol, aneurism and, iv, 476  
   in protein metabolism, ii, 584  
 Alcoholic neuritis, v, 579  
   trance, ii, 424  
 Alcoholism, ii, 396  
   acute hepatitis and, iii, 494  
   nephritis and, iii, 866  
   yellow atrophy of liver and, iii, 473  
   angioneurotic œdema and, iv, 999  
   arteriosclerosis and, iv, 455  
   carcinoma of stomach and, iii, 201  
   chronic, ii, 422  
     nephritis and, iii, 880, 891  
     pancreatitis and, iii, 627  
     peritonitis and, iii, 717  
   cirrhosis of liver and, iii, 508, 536  
   encephalitis and, v, 490  
   epilepsy and, v, 593, 596  
   fatty liver and, iii, 467  
   idiocy and, ii, 407  
   Korsakow's psychosis and, ii, 406, 427  
   lobar pneumonia and, i, 206  
   myocarditis and, iv, 112  
   periodic ii, 423  
   syphilitic myelitis and, v, 238  
   tuberculosis of peritoneum and, iii, 721  
   typhoid fever and, i, 159  
 Aleukemic lymphocytoma, iv, 748  
 Alexia, v, 54  
   in brain tumor, v, 333  
 Alimentary glycosuria, ii, 690  
   system in tuberculosis, i, 387  
   tract in cardiac insufficiency, iv, 204  
   in exophthalmic goitre, iv, 867  
   syphilis of, ii, 174  
   in typhoid fever, i, 82  
 Alimentary tract in uremia, iii, 837  
   in whooping cough, i, 660  
 Alkali-free diet, ii, 526  
 Alkapton substances, ii, 655  
 Alkaptonuria, ii, 655, 711; iii, 817  
 Allergic phenomena in man, i, 62  
 Allergy, immunity and, i, 65  
 Allochiria, v, 268  
   in tabes dorsalis, v, 838  
 Alloxuric bodies, ii, 732  
 Alopecia in exophthalmic goitre, iv, 867  
   in syphilis, ii, 167  
   from x-rays, ii, 336  
 Alternation of heart-beat, iv, 96  
 Alveolar sarcoma of brain, v, 314  
 Amaurosis, v, 34, 480  
   in anemia, iii, 836  
   in nephritis, iii, 865, 910  
 Amaurotic family idiocy, v, 863  
 Amblyopia, v, 341, 480  
   in lobar pneumonia, i, 263  
 Amboceptors, i, 54  
   in tuberculosis, i, 314  
 Ambulatory plague, i, 632  
   typhoid fever, i, 92, 153  
 Amimia in aphasia, v, 296  
 Amino-acids, ii, 506, 507, 581, 587, 644  
   in metabolism, ii, 654, 662  
 Amnesia, ii, 439; v, 53, 270  
 Amnesic aphasia, v, 54, 74, 298  
 Amoeba in carcinoma of stomach, iii, 213  
   Ehrenberg, ii, 22, 28  
   of human intestine, ii, 18  
   lobospinosa, ii, 24  
 Amebic colitis, ii, 17  
   dysentery, ii, 17  
     typhoid fever and, i, 158  
 Amphistoma hominis, ii, 238  
 Ampulla of Vater, carcinoma of, iii, 594  
 Amyl alcohol, ii, 400  
 Amyloid casts, iii, 799  
   degeneration, intestinal hemorrhage  
     and, iii, 307  
   of liver, iii, 469  
   of lymph nodes, iv, 746  
   of myocardium, iv, 116  
   syphilis and, ii, 152  
   disease, abscess of lung and, ii, 995  
   of intestines, iii, 392  
   in tuberculosis, i, 435  
   kidney, iii, 933  
   liver, ascites and, iii, 658  
   spleen, iv, 949  
   tumors of tongue, iii, 66  
 Amylopsin, iii, 289  
 Amyotonia congenita, v, 893  
 Amyotrophic lateral sclerosis, v, 89  
   syringomyelia, v, 220  
 Anabolism, ii, 549  
 Anacidity of stomach, iii, 153  
 Anacusis, v, 33  
 Analgesia, v, 30  
   dolorosa, v, 30  
   in hysteria, v, 682  
   in syringomyelia, v, 223  
 Anaphylactic shock, i, 730  
 Anaphylaxis, i, 61

- Anarthria, v, 74  
 Anchylostomiasis, ii, 271  
 Anemia in Addison's disease, iv, 791, 794  
   of adrenal glands, iv, 782  
   in amœbic dysentery, ii, 38, 68  
   in amyloid kidney, iii, 935  
   aplastic, iv, 644  
     purpura hemorrhagica and, iv, 699  
   of brain, v, 392  
   brick-makers', ii, 271  
   in carcinoma of intestines, iii, 430  
     of stomach, iii, 208  
   cryptogenic, iv, 619  
   gastric ulcer and, iii, 171, 176  
   in Hodgkin's disease, iv, 768  
   in hysteria, v, 688  
   of infancy, iv, 658  
   intestinal hemorrhage and, iii, 307  
   of kidney, iii, 787  
   in leukemia, iv, 665, 671  
   miners', ii, 271  
   in multiple neuritis, v, 585  
   myelophthisic, iv, 645  
   neuralgia and, v, 726  
   pernicious, iv, 619  
     pigmentation of liver and, iii, 471  
   posthemorrhagic, acute, iv, 655  
   in Raynaud's disease, iv, 983  
   in rickets, ii, 790  
   secondary, iv, 654  
     chronic, iv, 658  
     septic, acute, iv, 658  
     of spinal cord, v, 255  
   splenic, iv, 954, 957  
     secondary anemia and, iv, 658  
   symptomatic, iv, 654  
   in syphilis, ii, 151, 158  
   in tuberculosis, i, 433  
     of kidney, iii, 985  
   tunnel, ii, 271  
 Anergy, i, 62  
 Anesthesia, v, 28  
   in brain tumor, v, 337  
   in compression of spinal cord, v, 191  
   in hysteria, v, 680, 681  
   of larynx, v, 522  
   in migraine, v, 713, 714  
   in neuralgia, v, 728  
   in occupation neuroses, v, 747  
   in peripheral embolism, iv, 568  
     thrombosis, iv, 568  
   in Raynaud's disease, iv, 987  
   in syringomyelia, v, 223  
   in tabs dorsalis, v, 838  
 Aneurism, iv, 472  
   of aorta, abdominal, iv, 509  
     arch of, iv, 492  
     bronchostenosis and, ii, 921  
     descending thoracic, iv, 508  
     dilatation, iv, 484  
     syphilis and, ii, 183  
     thoracic, iv, 519  
   arteriovenous, iv, 472, 510  
     of abdominal aorta and inferior vena cava, iv, 513  
   Aneurism, arteriovenous, of aorta and pulmonary artery, iv, 513  
     and superior vena cava, iv, 512  
     internal, iv, 512  
     traumatic, iv, 510  
   atheroma and, iv, 477  
   blood-pressure and, iv, 478  
   bones and, iv, 482  
   of cerebral arteries, v, 398  
   circumscribed saccular, iv, 472  
   cirroid, iv, 486  
     of coronaries, iv, 437  
   cylindrical, iv, 479  
   dilatation, iv, 472, 484  
   dissecting, iv, 472, 487, 489  
     healed, iv, 490  
   of ductus arteriosus Botalli, iv, 419  
   embolism and, iv, 476, 557  
   erosion, iv, 472  
   false, iv, 472  
   fusiform, iv, 479  
   of heart, iv, 115, 127, 482  
   of hepatic artery, iii, 458  
   miliary, hematemeses from, iii, 283  
   mural, iv, 483  
   mycotic, iv, 476  
   parasitic, iv, 472  
   sacculated, iv, 479, 492  
   of sinus of Valsalva, iv, 492  
   true, iv, 472  
   of undefended space of interventricular septum, iv, 369  
   of valves of heart, iv, 482  
   varicose, iv, 510  
   without pulsation, iv, 518  
 Aneurismal pouching of fossa ovalis, iv, 359  
   varix, iv, 510  
 Anger, v, 61  
 Angina abdominis, iv, 464  
   Ludwig's, ii, 846; iii, 75  
   pectoris, iv, 289  
 Angioma of brain, v, 317  
   cavernosum of spleen, iv, 550  
   chyle, iv, 586  
   of lips, iii, 45  
   of liver, iii, 546  
   of peritoneum, iii, 733  
 Angioneuroses, purpura and, iv, 707  
 Angioneurotic group, iv, 707  
   cedema, iv, 998  
     of mouth, iii, 55  
     of pharynx, ii, 847  
 Anguillula aceti, ii, 315  
 Anguilluliasis, ii, 285  
 Anguillulina putrefaciens, ii, 294  
 Animal parasites, ii, 217  
 Anisocoria in aneurism of aorta, iv, 494  
 Ankylostomiasis, ii, 271  
 Annular rickets, v, 955  
 Annulus ovalis, iv, 326  
 Anomalies of adrenal glands, iv, 781  
   of arch of aorta, iv, 433  
   of auriculoventricular cusps, iv, 407  
   of biliary ducts, iii, 547  
   of conduct; v, 63

- Anomalies of coronary arteries, iv, 437  
 of ductus arteriosus Botalli, iv, 409  
 of gall-bladder, iii, 547  
 of ideas, v, 52  
 of heart as a whole, iv, 350  
 of kidney, iii, 785  
 of pericardium, iv, 344  
 of pulmonary arteries, iv, 439  
   veins, iv, 441  
 of reflexes, v, 76  
 of semilunar cusps, iv, 400  
 of sense of pain, v, 29  
 of septum of heart, iv, 352  
 of systemic veins, iv, 439  
 of thoracic duct, iv, 580  
 of thymus, iv, 908  
 of tongue, iii, 60  
 of will, v, 63
- Anomalous chordæ tendineæ, iv, 354  
   septa in left auricle, iv, 352  
   in right auricle, iv, 352  
   in ventricles, iv, 354
- Anomia, v, 76
- Anopsia, v, 34
- Anosmia, v, 33, 471
- Anthraxemia, i, 639
- Anthraxosis, ii, 976  
   hepatic, iii, 470
- Anthrax, i, 639
- Anti-anaphylaxis, i, 62
- Antibodies, i, 46, 47, 49, 50, 51, 313
- Antigen-antibody reactions, i, 51
- Antigens, i, 48
- Antiperistalsis of intestines, iii, 287, 288
- Antitoxins, i, 47, 53  
   in diphtheria, i, 54, 715  
   in tuberculosis, i, 313
- Antituberculins in tuberculosis, i, 313
- Antityphoid inoculation, i, 176
- Anuria, iii, 841
- Aorta, abdominal, aneurism of, iv, 509  
   aneurism of, iv, 492, 493, 508  
     bronchostenosis and, ii, 921  
     syphilis and, ii, 183  
   arch of, aneurism of, iv, 492  
     anomalies of, iv, 433  
     ascending, aneurism of, iv, 493  
     double, iv, 434  
     right, iv, 434  
     transverse, aneurism of, iv, 493  
   atresia of, congenital, iv, 399  
   coarctation of, iv, 420  
     endocarditis and, iv, 429  
   descending thoracic, aneurism of, iv, 508  
     right subclavian artery from, iv, 436  
   dilatation of, iv, 484  
     dynamic, iv, 513  
   elasticity of, iv, 34  
   embolism of, iv, 565  
   hypoplasia of, iv, 431  
     aneurism and, iv, 478  
     defects of interauricular septum and, iv, 359  
   intima of, fissure of, iv, 487  
   rupture of, iv, 487
- Aorta, rupture of, in coarctation, iv, 426  
   stenosis of, congenital, iv, 399  
   thrombosis of, iv, 565
- Aortic arches, primitive, development of, iv, 329  
   atresia, congenital, iv, 398  
   insufficiency, iv, 217  
   septum, defects of, iv, 374  
     development of, iv, 327  
     deviation of, iv, 379  
   stenosis, iv, 228
- Aortitis, acute, iv, 450  
   chronic, circulation in, iv, 25  
   in pneumonia, iv, 450  
   in rheumatic fever, iv, 450, 475  
   syphilitic, ii, 182; iv, 474  
   tuberculous, iv, 452
- Aphasia, v, 292  
   in abscess of brain, v, 381  
   in apoplexy, v, 457  
   in arteriosclerosis, iv, 461; v, 397  
   in brain tumor, v, 331  
   Broca's, v, 295  
   motor, v, 75, 292, 295  
   optic, v, 54, 299  
   in pachymeningitis interna, v, 165  
   sensory, v, 75, 292, 296  
   subcortical, v, 75  
   visual, v, 298
- Aphemia, v, 300
- Aphthæ, Bednar's, iii, 52
- Apthous fever, i, 1025  
   stomatitis, iii, 46
- Apical pneumonia, i, 236
- Aplasia, myeloplastic, v, 978
- Aplastic anemia, iv, 644
- Apoplexy, v, 405  
   arteriosclerosis and, v, 396, 427  
   brain tumor and, v, 419  
   cerebral aneurism and, v, 399, 429  
     diplegia and, v, 419  
     hemorrhages and, v, 410, 411, 414, 415  
     palsies and, v, 410  
   chronic nephritis and, v, 427  
   diabetes mellitus and, v, 427  
   diphtheria and, v, 417  
   embolism and, v, 418, 421, 424, 425, 427  
   hemiplegia and, v, 409, 412, 457  
   hemorrhages and, v, 422, 424, 427, 428, 431  
   hysteria and, v, 419  
   measles and, v, 417  
   pancreatic, iii, 610  
   porencephaly and, v, 409  
   pregnancy and, v, 423, 425  
   rupture of aneurism and, v, 429  
   scarlet fever and, v, 417  
   syphilis and, v, 423, 425, 426, 427  
   thrombosis and, v, 423, 425  
   typhoid fever and, v, 417, 425
- Appendicitis, iii, 355  
   abscess of liver and, iii, 497  
   in amœbic dysentery, ii, 44  
   catarrhal, iii, 372  
   cholelithiasis and, iii, 571

- Appendicitis, chronic, iii, 365, 375  
   gangrenous, iii, 372  
   hyperacidity and, iii, 114  
   kidney infections and, iii, 952  
   lymphoid tissue and, iii, 361  
   mesenteric lymphadenitis and, iv, 736  
   mild, iii, 373  
   myocarditis and, iv, 113  
   nephroptosis and, iii, 757  
   parasites and, iii, 360  
   rheumatic fever and, i, 998  
   simple, iii, 372  
   subphrenic abscess and, iii, 700  
   thrombosis of portal vein and, iv, 544  
   in tuberculosis, i, 432  
   typhoid fever and, i, 159  
 Appendicular abscess, pneumothorax and, iii, 711  
   colic, iii, 373  
 Appendix, actinomycosis of, iii, 386  
   cystic distension of, iii, 386  
   gangrene of, iii, 363  
   inflammation of, iii, 355  
   mesentery of, iii, 357  
   necrosis of, iii, 363  
   obliteration of, iii, 366  
   palpation of, iii, 371  
   polyps of, iii, 386  
   sarcoma of, iii, 387  
   tuberculosis of, iii, 385  
   tumors of, iii, 386  
 Apraxia, v, 66, 75, 269  
   aphasia and, 299  
   in diseases of corpus collosum, v, 276  
 Aptyalism, iii, 68  
 Arachnoidal cysts of brain, v, 318  
 Aran-Duchenne muscular atrophy, v, 80  
   paralysis in lead poisoning, ii, 366  
 Ardent fever, i, 1011  
 Argas miniatus, ii, 317  
   persicus, ii, 317  
   reflexus, ii, 317  
 Argopsylla gallinacea, ii, 325  
 Argyll-Robertson pupil, v, 487  
   in general paresis, v, 856  
   in tabes dorsalis, v, 836  
 Argyria, ii, 384  
 Arrhythmia, cardiac, iv, 29  
   in exophthalmic goitre, iv, 861  
   in lobar pneumonia, i, 256  
   in myocarditis, iv, 133  
   in syphilis of myocardium, iv, 143  
 Arm in aneurism of aorta, iv, 495  
   paralysis of, v, 551, 552  
 Arrow-headed eelworms, ii, 288  
 Arsenic poisoning, ii, 373  
 Arterial spasm, gastric ulcer and, iii, 171  
   thrombosis, iv, 537  
   trunk, common, iv, 375  
   trunks, transposition of, iv, 379  
 Arteries in aortic insufficiency, iv, 223, 226  
   in arteriosclerosis, iv, 466  
   carotid, embolism of, iv, 567  
   thrombosis of, iv, 567  
   cerebral, aneurism of, v, 398  
   obliteration of, v, 400  
   rupture of, v, 400  
 Arteries, cerebral, thrombosis of, v, 400  
   in chronic nephritis, iii, 898, 903  
   coronary, in arteriosclerosis, iv, 463  
   embolism of, iv, 569  
   thrombosis of, iv, 569  
   gumma of, ii, 181  
   hepatic, embolism of, iv, 569  
   peripheral, embolism of, iv, 568  
   thrombosis of, iv, 568  
   pulmonary, dilatation of, iv, 398  
   renal, anomalies of, iii, 786  
   retinal, embolism of, iv, 570  
   thrombosis of, iv, 570  
   in rheumatic fever, i, 989  
   subclavian, embolism of, iv, 567  
   thrombosis of, iv, 567  
   syphilis of, ii, 181  
   in typhoid fever, i, 81, 127  
 Arteriosclerosis, iv, 452  
   aortic insufficiency and, iv, 218  
   stenosis and, iv, 218  
   apoplexy and, v, 427  
   of brain, v, 393  
   cardiac insufficiency and, iv, 199  
   cerebral aneurism and, v, 399  
   thrombosis and, v, 400  
   coronary thrombosis and, iv, 569  
   diabetes mellitus and, ii, 682  
   experimental, iv, 458  
   in gout, ii, 742  
   hyperpietic, iv, 457  
   interstitial nephritis and, ii, 893  
   of intestines, iii, 437  
   lead poisoning and, ii, 365; iv, 455  
   mitral insufficiency and, iv, 232  
   neuralgia and, v, 727  
   in Paget's disease, v, 973  
   peritonitis and, iii, 716  
   Raynaud's disease and, iv, 978, 990  
   Stokes-Adams' syndrome and, iv, 306  
   syphilis and, ii, 162; iv, 454  
   toxic, iv, 457  
 Arteriosclerotic kidney, iv, 463  
 Arteriovenous aneurism, iv, 472, 510  
 Arteritis, iv, 449  
 Arthracotic cirrhosis of liver, iii, 531  
 Arthritis, acute endocarditis and, iv, 155  
   in cerebrospinal meningitis, i, 610  
   in chickenpox, i, 853  
   deformans, v, 895  
   atrophic, v, 920  
   gout and, v, 930  
   hypertrophic, v, 920  
   infections and, v, 900, 902, 903  
   influenza and, v, 901  
   metabolism and, v, 903, 907  
   monarticular, v, 919, 934  
   peri-articular, v, 908  
   Raynaud's disease and, v, 929  
   scleroderma and, v, 929  
   of special joints, v, 910  
   Still's disease and, v, 917, 931  
   gonorrhoeal, i, 753, 756  
   in hemophilia, iv, 724  
   infectious, v, 896  
   in lobar pneumonia, i, 263  
   in measles, i, 907



- Arthritis in osteo-arthropathy, v, 985  
 . in purpura hemorrhagica, iv, 712  
 in rheumatic fever, i, 990  
 in scarlet fever, i, 878  
 in scleroderma, iv, 1018, 1021  
 in tuberculosis, i, 504  
 in typhoid fever, i, 148  
 villous, v, 929
- Arthropathy in syringomyelia, v, 225  
 in tabes dorsalis, v, 842
- Ascariasis, ii, 287
- Ascaris lumbricoides, ii, 287  
 appendicitis and, iii, 360  
 maritima, ii, 288  
 texana, ii, 288
- Ascending aorta, aneurism of, iv, 493  
 multiple neuritis, v, 576
- Ascites, iii, 657  
 chyloform, iii, 661  
 chylous, iii, 660; iv, 590  
 fatty, iii, 661  
 lactescent, iii, 662  
 milky, iii, 662  
 non-fatty, iii, 662
- Ascitic fluid, iii, 659, 665
- Asepsis in genito-urinary diagnosis, iii, 1029
- Ash-free diet, ii, 526
- Asiatic cholera, i, 672  
 clonorchiasis, ii, 230
- Aspergillosis, pulmonary, i, 1058
- Aspergillus fumigatus, i, 1058
- Asphyxia, cardiac insufficiency and, iv, 195  
 in enlargement of thymus, iv, 916  
 mechanics of, ii, 835
- Aspiration pneumonia, i, 206; ii, 958
- Association tests, v, 48
- Astasia-abasia, v, 815  
 hysteria and, v, 816
- Astereognosis, v, 30, 268  
 in lesion of optic thalamus, v, 281  
 in tabes dorsalis, v, 838
- Asthenic paralysis, v, 798  
 pneumonia, i, 236
- Asthenopia, v, 36
- Asthma, ii, 931  
 cardiac, iv, 207  
 gout and, ii, 933  
 hay, ii, 940  
 Kopp's, iv, 918  
 pregnancy and, ii, 933  
 renal, ii, 905  
 thymic, iv, 918  
 in tuberculosis, i, 430
- Astigmatism, v, 36
- Asyllabia, v, 298
- Asymbolia, v, 54, 269
- Asynergy in brain abscess, v, 381  
 tumor, v, 339  
 in lesion of cerebellum, v, 286
- Asystole, ventricular, iv, 89
- Ataxia, v, 71  
 cerebellar, v, 72  
 Marie's hereditary, v, 130, 136  
 Friedreich's, v, 130
- Atelectasis in aneurism of aorta, iv, 503  
 in lobar pneumonia, i, 248  
 tumors of lung and, ii, 1001
- Ateleiosis, iv, 817
- Atheroma, iv, 432  
 aneurism and, iv, 477  
 in coarctation of aorta, iv, 426  
 of coronary arteries, cardiac insufficiency and, iv, 195
- Athetoid movements, v, 70
- Athetosis in cerebral diplegia, v, 420  
 in lesion of optic thalamus, v, 280
- Athymia, v, 61
- Athyrea, iv, 880
- Athyreosis, iv, 880
- Athyroidism, iv, 880
- Atonic constipation, iii, 313
- Atony of muscle, v, 68  
 of stomach, iii, 160
- Atresia of aorta, congenital, iv, 398, 399  
 mitral, iv, 406  
 pulmonary, iv, 388  
 tricuspid, iv, 403
- Atrophic arthritis deformans, v, 920  
 diffuse scleroderma, iv, 1016
- Atrophy of adrenal glands, iv, 788  
 of cerebellum, v, 287  
 in erythromelalgia, iv, 1028  
 facial, in migraine, v, 719  
 gastric, pernicious anemia and, iv, 623  
 of liver, iii, 465  
 acute yellow, iii, 472  
 muscular, v, 68  
 in arthritis deformans, v, 914  
 in diseases of peripheral nerves, v, 534  
 progressive, Charcot-Marie-Tooth  
 type, v, 109  
 neural, v, 109  
 spinal, v, 80  
 of childhood, v, 87  
 type Duchenne-Aran, v, 80  
 Werdnig-Hoffmann, v, 87
- olivopontocerebellar, v, 287  
 optic, v, 37, 478  
 in osteo-arthropathy, v, 986  
 of salivary gland, iii, 74  
 in scleroderma, iv, 1019  
 of thymus, iv, 910  
 of tongue, iii, 66; v, 531
- Attention, anomalies of, v, 45
- Attonnity, v, 65
- Auditory nerve, diseases of, v, 506  
 vertigo, v, 512
- Aura in epilepsy, v, 600
- Aural vertigo, v, 513
- Auricle, left, double, iv, 352  
 hypertrophy of, iv, 185  
 right, hypertrophy of, iv, 184
- Auricles in circulation, role of, iv, 24
- Auricular fibrillation, iv, 86  
 flutter, iv, 86
- Auriculo-ventricular bundle, iv, 80  
 cusps, anomalies of, iv, 407  
 development of, iv, 328  
 orifice, defects of, iv, 403  
 double, iv, 407
- Auscultation in lobar pneumonia, i, 244
- Auto-inoculation in carcinoma of stomach, iii, 201

Auto-intoxication, ii, 503  
 Autolysis, ii, 663  
 Automatic mechanism in intestinal processes, iii, 291  
 Automatism, ii, 424; v, 44  
 Autonomic system, iii, 24, 35, 106  
 Autopsyche, v, 43  
 Axillary glands, lymphadenitis of, iv, 735  
   tuberculosis of, iv, 744  
 Azotorrhœa in calculus of pancreas, iii, 635  
   in disease of pancreas, iii, 602

## B

BABINSKI'S hypothesis of hysteria, v, 658  
 sign, v, 78

  in apoplexy, v, 440  
   in cerebrospinal meningitis, i, 606  
   in disseminated sclerosis, v, 149  
 Bacillary dysentery, i, 766  
 Bacillen emulsion, i, 305  
 Bacilluria in typhoid fever, i, 142  
 Bacillus *aërogenes capsulatus* in acute  
   peritonitis, iii, 676  
   appendicitis, iii, 361  
   in intestines, iii, 296  
   anthracis, i, 561, 641  
   *bifidus communis* in intestines, iii, 296  
 Boas-Oppler, iii, 212  
 botulinus, ii, 459, 460, 469  
 coli in appendicitis, iii, 361  
   in diarrhœa, iii, 325  
   in fat necrosis, iii, 607  
   in intestines, iii, 296  
   in jaundice, iii, 448  
   in peritonitis, iii, 674, 717  
 diphtheria, i, 689  
   in mouth, iii, 40  
 dysenteria, i, 766  
 enteritidis, ii, 454, 459, 463, 464  
   in diarrhœa, iii, 325  
 icteroides, i, 937  
 influenza in bronchopneumonia, ii, 959  
 lactis *aërogenes* in appendicitis, iii, 361  
   in intestines, iii, 296  
 lepræ, i, 523  
   in mouth, iii, 41  
 mallei, i, 649  
 maximus in mouth, iii, 40  
 necrodenitis in mouth, iii, 40  
 paratyphosus, ii, 459  
 pertussis, i, 656  
 pestis, i, 623  
 piscicidus, ii, 452  
 proteus fluorescens, i, 1015  
   in jaundice, iii, 448  
 pseudodiphtheria in mouth, iii, 40  
 putrificus in intestines, iii, 296  
 pyocyaneus, i, 558  
   in acute peritonitis, iii, 675  
   in appendicitis, iii, 361  
   in diarrhœa, iii, 325  
   in mouth, iii, 40  
 salmonicida, ii, 452  
 tetani, i, 575  
 tuberculosis, i, 294

Bacillus tuberculosis in mouth, iii, 41  
   typhosus, i, 77  
   in peritonitis, iii, 675, 717  
 Bacteria in acute enteritis, iii, 329  
   in constipation, iii, 312  
   in intestines, iii, 295  
   multiple neuritis and, v, 575, 576  
   myelitis and, v, 229  
   of normal air passages, ii, 883  
   pathogenic, i, 18  
   in urethra, iii, 943  
 Bacteriology of acute cholecystitis, iii, 561  
   peritonitis, iii, 674  
   of appendicitis, iii, 361  
   of ascites, iii, 660  
   of biliary infections, iii, 549  
   of bronchopneumonia, ii, 959  
   of cholelithiasis, iii, 570  
   of chorea, v, 778  
   of cystitis, iii, 944  
   of diarrhœa, iii, 324  
   of encephalitis, v, 362  
   of gangrenous stomatitis, iii, 49  
   of infections of kidney, iii, 947, 950  
   of infective myelitis, v, 231  
   of intestinal tract, iii, 295  
   of Landry's paralysis, v, 248  
   of mouth, iii, 39  
   of mumps, i, 919  
   of purpura, iv, 697  
   of pyelitis, iii, 947, 950  
   of rheumatic fever, i, 975  
   of serofibrinous pleuritis, ii, 1032  
   of subphrenic abscess, iii, 706  
   of suppurative pancreatitis, iii, 623  
   of tuberculosis of peritoneum, iii, 721  
   of ulcerative stomatitis, iii, 48  
   of urethritis, iii, 943, 946  
   of urine, iii, 943  
 Bacteriolysins in tuberculosis, i, 314  
 Bacterioproteins, i, 41  
 Bacteriotoxins, i, 34  
 Bacterium gingivitis in mouth, iii, 40, 55  
 Bacteriuria, iii, 948  
 Baelz's disease, iii, 45  
 Bands from chronic peritonitis, iii, 715  
 Barbadoes leg, iv, 596  
 Basal ganglia, tumors of, v, 334  
 Basic meningitis, posterior, i, 615  
 Bathyanesthesia, v, 30  
 Bathyesthesia, v, 17  
 Becquerel's rays, ii, 339  
 Bed-bugs, ii, 327  
 Bednar's aphthæ, iii, 52  
 Bence-Jones proteins in urine, iii, 819  
 Benedikt's syndrome, v, 369  
 Beriberi, ii, 489  
 Bertiella satyri, ii, 246, 255  
 Bifid apex of heart, iv, 350  
 Bile, iii, 289  
 Bilharziosis, ii, 239  
 Biliary cirrhosis of liver, iii, 535  
   ducts, abscess of liver and, iii, 498  
   anomalies of, iii, 547  
   carcinoma of, iii, 593  
   common, obstruction of, iii, 581  
   congenital obliteration of, iii, 547

- Biliary ducts, extrahepatic, carcinoma of, iii, 594  
 fistula in cholelithiasis, iii, 584  
 tract, digestion and, iii, 34  
 infections of, iii, 548
- Bilirubin in liver, iii, 470
- Biliverdin in liver, iii, 470
- Biological hypothermia of hysteria, v, 665
- Biot's respiration, ii, 835
- Birth palsies, brachial, v, 552  
 traumatism, epilepsy and, v, 594
- Black tongue, iii, 61
- Blackwater fever, ii, 109
- Bladder in hemiplegia, v, 467  
 in renal calculus, iii, 1021  
 rupture of peri-appendicular abscess into, iii, 365  
 in syphilitic myelitis, v, 240  
 in tabes dorsalis, v, 840  
 tuberculosis of, i, 366  
 kidney and, iii, 981  
 in typhoid fever, i, 86
- Blastomycetes, i, 1054
- Blastomycosis, i, 1054  
 of myocardium, iv, 144
- Bleeding in apoplexy, v, 453  
 in hysteria, v, 687  
 in purpura, iv, 696
- Blind spot, v, 34
- Blindness, v, 34  
 in amaurotic family idiocy, v, 869  
 cortical, v, 54  
 day, v, 480  
 in hysteria, v, 684  
 in lead poisoning, ii, 368  
 mind, v, 53, 271  
 night, v, 480  
 psychic, v, 299  
 in retinal embolism, iv, 571  
 thrombosis, iv, 571  
 smallpox and, i, 812  
 in uremia, iii, 836  
 word, v, 54
- Blood in abscess of liver, iii, 502  
 in acromegaly, iv, 812  
 in acute nephritis, iii, 864  
 peritonitis, iii, 683  
 yellow atrophy of liver, iii, 476  
 in Addison's disease, iv, 793  
 in aplastic anemia, iv, 645  
 in arthritis deformans, v, 916  
 in atypical leukemia, iv, 679  
 bacteriology of, i, 559  
 in blackwater fever, ii, 115  
 in carcinoma of stomach, iii, 204  
 cells, enumeration of, iv, 606  
 in cerebrospinal meningitis, i, 605  
 in chloroma, iv, 682  
 in chlorosis, iv, 651  
 in cirrhosis of liver, iii, 525, 539  
 coagulation of, iv, 719  
 color index of, iv, 607  
 congenital cyanosis and, iv, 335  
 cultures, i, 559  
 in diabetes mellitus, ii, 695  
 distribution of, in circulatory diseases, iv, 18
- Blood in echinococcus disease of pleura, ii, 1068  
 in enlargement of thymus, iv, 924  
 in exophthalmic goitre, iv, 867  
 in feces in amoebic dysentery, ii, 36  
 formation of, iv, 603  
 in gonorrhœal arthritis, i, 757  
 in gout, ii, 742  
 in Hodgkin's disease, iv, 768  
 in hysteria, v, 688  
 in infantile scurvy, ii, 806  
 in infectious diseases, i, 42  
 in influenza, i, 545  
 in jaundice, iii, 452  
 in kala-azar, iv, 972  
 in lobar pneumonia, i, 218, 234  
 in lymphoid leukemia, iv, 672  
 in malarial fever, ii, 88  
 mass, movement of, iv, 18  
 in measles, i, 905  
 in myeloid leukemia, iv, 666  
 in myxœdema, iv, 894  
 in obesity, ii, 765, 767  
 "occult" tests for, iii, 180  
 in osteomalacia, v, 952  
 in pellagra, ii, 484  
 in pernicious anemia, iv, 635, 639  
 in plague, i, 631  
 platelets, counting of, iv, 694  
 in pleuritis, ii, 1009, 1024, 1043  
 in polycythemia, iv, 685  
 portal, spleen and, iv, 936  
 in posthemorrhagic anemia, iv, 656  
 in pseudoleukemia, iv, 681  
 in purpura, iv, 693, 699  
 renal function and, iii, 770  
 in rheumatic fever, i, 989  
 in rubella, i, 914  
 in scarlet fever, i, 876  
 in scleroderma, iv, 1021  
 in smallpox, i, 808  
 in sporadic cretinism, iv, 885  
 in syphilis, ii, 151  
 in toxemia, i, 568  
 in tuberculosis, i, 382  
 of peritoneum, iii, 726  
 in typhoid fever, i, 130  
 in uremia, iii, 831  
 in urine, iii, 802  
 in whooping cough, i, 659
- Blood-fluke infection, ii, 239
- Blood-making organs, iv, 611  
 in infectious diseases, i, 42
- Blood-pressure in Addison's disease, iv, 793  
 in aneurism of aorta, iv, 498  
 arteriosclerosis and, iv, 456  
 in cardiac insufficiency, iv, 211  
 high, aneurism and, iv, 478  
 muscular work and, iv, 456  
 in hypertrophy of heart, iv, 186  
 in lobar pneumonia, i, 226  
 in mitral insufficiency, iv, 235  
 in myocarditis, iv, 121  
 in nephritis, iii, 903, 904, 921, 927  
 in patent ductus arteriosus, iv, 411  
 in tuberculosis, i, 385

- Blood-pressure in typhoid fever, i, 127  
 Bloodvessels in aneurism of aorta, iv, 504  
   in chronic nephritis, iii, 884, 896  
   congenital cyanosis and, iv, 335  
   in goitre, iv, 843  
   in jaundice, iii, 453  
   syphilis of, ii, 180  
   in syphilitic myelitis, v, 238  
   tuberculosis of, i, 360, 386  
 Blue line in lead poisoning, ii, 363  
 Boas-Oppler bacillus, iii, 212  
 Boils in diabetes mellitus, ii, 704  
 Bone lesions, gonococcal, i, 760  
   in typhoid fever, i, 146  
 Bone-marrow, iv, 611  
   in anemia, iv, 612, 645  
   in blackwater fever, ii, 113  
   in Gaucher's splenomegaly, iv, 963  
   in Hodgkin's disease, iv, 764  
   hyperplastic, iv, 612  
   in kala-azar, iv, 970  
   in malarial fever, ii, 72  
   in myeloid leukemia, iv, 663  
   in pernicious anemia, iv, 623, 625  
   in scarlet fever, i, 866  
   in smallpox, i, 790  
   in splenic anemia, iv, 956  
   tuberculosis of, i, 371  
 Bones in achondroplasia, v, 956  
   in acromegaly, iv, 807  
   aneurism and, iv, 482  
   in arthritis deformans, v, 904, 906, 907  
   in creatinism, iv, 881, 884  
   in exophthalmic goitre, iv, 868  
   in facial hemiatrophy, v, 992  
   in hypopituitarism, iv, 815  
   in myositis ossificans, v, 883, 884  
   in osteogenesis imperfecta, v, 979  
   in osteomalacia, v, 949  
   in Paget's disease, v, 971  
   in scleroderma, iv, 1020  
   in syringomyelia, v, 225  
 Boophilus annulatus, ii, 317  
 Botryoid liver, ii, 177  
 Botulismus, ii, 460  
 Bovovaccines, i, 305  
 Bowels, ulceration of, iii, 343  
 Brachial neuralgia, v, 737  
   neuritis, v, 553  
   plexus, paralysis of, v, 550  
 Brachiocephalic trunk, common, iv, 437  
 Bradycardia, iv, 282  
   in hysteria, iv, 283  
   in intoxications, iv, 285  
   in jaundice, iii, 453  
   in melancholia, iv, 283  
   myocardial, iv, 282  
   in neurasthenia, iv, 283  
   in pericarditis, iv, 283  
   simple, iv, 82  
   in Stokes-Adams syndrome, iv, 305  
   in tabes dorsalis, iv, 283  
   tachycardia and, iv, 302  
   vagus and, iv, 282, 284  
 Bradylalia, v, 74  
 Brain, abscess of, v, 378  
   in bronchiectasis, ii, 917  
 Brain, abscess of, traumatic, v, 382  
   in acromegaly, iv, 808  
   actinomycoma of, v, 32  
   acute red softening of, v, 359  
   in alcoholism, ii, 401  
   in amaurotic family idiocy, v, 864  
   anemia of, v, 392  
   angioma of, v, 317  
   arteries of, aneurism of, v, 398  
     embolism of, v, 400  
     rupture of, v, 400  
     thrombosis of, v, 400  
   arteriosclerosis of, v, 393  
   in blackwater fever, ii, 113  
   bloodvessels of, diseases of, v, 387  
   carcinoma of, v, 315  
   centre of hearing in, v, 271  
     of smell in, v, 271  
   centrum semiovale, lesion of, v, 275  
   cerebellum, lesion of, v, 284  
   in cerebrospinal meningitis, i, 597  
   chloroma of, v, 315  
   cholesteatoma of, v, 316  
   chordoma of, v, 317  
   convulsions of, lesions of, v, 274  
   corpora striata, lesion of, v, 277  
   corpus callosum, lesion of, v, 276  
   cortex of, motor area of, v, 265, 273  
     sensory area of, v, 265, 273  
   crura cerebri, lesion of, v, 281  
   cysticercus cellulose of, v, 318  
   cystoglioma of, v, 313  
   cystosarcoma of, v, 315  
   cysts of, v, 318  
   in delirium tremens, ii, 405  
   enchondroma of, v, 317  
   endothelioma of, v, 313  
   epithelioma of, v, 315  
   fibroma of, v, 317  
   glioma of, v, 311  
   granuloma of, infectious, v, 319  
   hyperemia of, v, 392  
   inflammation of, v, 359  
   injuries of, in apoplexy, v, 438  
   internal capsule, lesion of, v, 276  
   lenticular nucleus, lesion of, v, 278  
     zone, lesion of, v, 278  
   lipoma of, v, 317  
   in malarial fevers, ii, 70  
   myeloma of, v, 315  
   myxosarcoma of, v, 315  
   neuroblastoma of, v, 317  
   neurocystoma of, v, 317  
   neurofibroma of, v, 314  
   neuroglioma of, v, 311  
   neuroma of, v, 317  
   optic thalamus, lesion of, 279  
   osteosarcoma of, v, 314  
   papilloma of, v, 315  
   in pellagra, ii, 477  
   pons, lesion of, v, 289  
   pontomedullo-cerebellar space, lesion  
     of, v, 289  
   psammoma of, v, 318  
   psammoma of, v, 315  
   quadrigeminal bodies, lesion of, v, 282  
   in Raynaud's disease, iv, 988

- Brain, sarcoma of, v, 313, 314  
 sclerosis of, v, 140  
 stem, tumors of, v, 335  
 syphilis of, v, 823  
 syphiloma of, v, 320, 321  
 tegmentum, lesion of, v, 281  
 teratoma of, v, 316  
 tuberculoma of, v, 319  
 tuberculosis of, i, 370  
 tuberculous meningo-encephalitis of, v, 319  
 tumors of, v, 308  
   in basal ganglia, v, 334  
   in brain stem, v, 335  
   in cavum Meckelii, v, 340  
   in centrum semiovale, v, 334  
   in cerebellum, v, 338  
   in corpora quadrigemina, v, 336  
   in corpus collosum, v, 335  
   in cranial base, v, 340  
   in crura, v, 335  
   dermoid, v, 316  
   in fourth ventricle, v, 338  
   in frontal lobe, v, 331  
   hemiplegia and, v, 419  
   in hypophysis cerebri, v, 341  
   in medulla oblongata, v, 337  
   migraine and, v, 720  
   in occipital lobe, v, 334  
   in paracentral convolutions, v, 332  
   in parietal lobe, v, 333  
   in pineal gland, v, 336  
   in pons, v, 336  
   in tegmentum, v, 336  
   in temporal lobe, v, 331  
   trauma and, v, 309  
   wet, ii, 415  
 Brass poisoning, ii, 386  
 Braunia jassyensis, ii, 246, 258  
 Break-bone fever, i, 950  
 Breast, tuberculosis of, i, 368  
 Breath, offensive, iii, 42  
 Brick-makers' anemia, ii, 271  
 Bright's disease, iii, 778, 845  
 Broad tapeworm, ii, 255  
 Broca's aphasia, v, 75, 295  
 Bromides in epilepsy, v, 620  
 Bronchi in aneurism of aorta, iv, 502, 507  
   foreign bodies in, ii, 926, 928  
   leprosy of, ii, 906  
   stenosis of, ii, 820  
   syphilis of, ii, 170, 906  
   tuberculosis of, i, 348; ii, 905  
 Bronchial calculi, ii, 925  
   glands in lobar pneumonia, i, 253  
   lymphadenitis of, iv, 735  
   tuberculosis of, iv, 743  
   obstruction, ii, 920  
 Bronchiectasis, ii, 913  
   abscess of brain and, v, 379  
   in aneurism of aorta, iv, 503  
   in cerebrospinal meningitis, i, 599  
   osteo-arthropathy and, v, 982  
   pneumokoniosis and, ii, 977  
   pneumothorax and, ii, 1074  
   in syphilis of bronchi, ii, 907  
 Bronchiectasis in syphilis of trachea, ii, 907  
   tricuspid insufficiency and, iv, 256  
   in tuberculosis, i, 430  
   in whooping cough, i, 660  
 Bronchitis, acute, ii, 881  
   albuminuric, ii, 904  
   bronchopneumonia and, ii, 887  
   capillary, ii, 887, 957  
   chronic, ii, 894  
   fetid, ii, 911  
   fibrinous, ii, 907  
   gangrenous, ii, 911  
   in gout, ii, 749  
   in influenza, i, 537, 540; ii, 902  
   in lobar pneumonia, i, 248  
   in malarial fevers, ii, 86  
   in measles, i, 906; ii, 901  
   in mitral insufficiency, ii, 903; iv, 236  
   stenosis, ii, 903  
   in nephritis, iii, 870, 905  
   plastic, ii, 907  
   pneumokoniosis and, ii, 977  
   pseudomembranous, ii, 907  
   purulent, ii, 897  
   putrid, ii, 911  
   in rickets, ii, 790  
   secondary, ii, 901  
   serous, ii, 896  
   tricuspid insufficiency and, iv, 246  
   in tuberculosis, i, 322, 419, 429  
   in typhoid fever, i, 121  
   in whooping cough, i, 660; ii, 902  
 Bronchoblennorrhœa, ii, 897  
 Bronchocele, iv, 835  
 Broncholiths, ii, 925  
 Bronchopneumonia, ii, 957  
   in bronchiectasis, ii, 917  
   in diphtheria, i, 693, 701; ii, 958  
   in fetid bronchitis, ii, 912  
   in influenza, i, 537, 540  
   in measles, i, 906; ii, 958, 963  
   pleuritis and, ii, 964, 1012  
   in scarlet fever, i, 864; ii, 958  
   in tuberculosis, i, 430; ii, 964  
   tuberculous, acute, i, 425  
   in typhoid fever, i, 122  
   in typhus fever, i, 930  
   in whooping cough, i, 660; ii, 958, 963  
 Bronchopulmonary lithiasis, ii, 925  
 Bronchostenosis, ii, 920  
 Bronze diabetes, ii, 681; iii, 529  
 Brown induration of lungs, ii, 945  
 Brown-Séquard paralysis in lesions of spinal cord, v, 185, 204  
 Brudzinski's reflex, i, 607  
 Bubo, iv, 738  
   gonorrhœal, iv, 739  
   indolent, iv, 740  
   in plague, i, 624  
 Bubonic plague, i, 632  
 Bulbar changes in amyotrophic lateral sclerosis, v, 91, 100  
 dyspnœa, iv, 40  
 measles in myasthenia gravis, v, 802  
 palsy, acute inflammatory, v, 368  
 progressive, v, 113

Bulbar paralysis, ascending, v, 798  
 Bulbus cordis, development of, iv, 325  
 Bulimia, iii, 132  
 Bundle of His, iv, 80, 203  
     development of, iv, 328  
     interventricular defects and, iv, 367  
     Stokes-Adams syndrome and, iv, 308  
 Burns from electricity, ii, 342  
     from heat, ii, 348  
     nephritis and, iii, 853  
 Burrowing fleas, ii, 325  
 Bursæ, tuberculosis of, i, 373  
 Butyl alcohol, ii, 400  
 Butyric acid in carcinoma of stomach, iii, 211

## C

CACHECTIC splanchnoptosis, iii, 754  
 Cachexia hypophyseopriva, v, 341  
     of malarial fever, ii, 88  
     miners', ii, 271  
     multiple sclerosis and, v, 576  
     thyroidea, iv, 888  
     in tuberculosis of kidney, iii, 985  
 Caisson disease, ii, 345  
 Calcareous degeneration of myocardium, iv, 116  
 Calcification of lymph nodes, iv, 746  
     of thymus, iv, 912  
 Calcium carbonate calculus, iii, 1009  
     metabolism of, ii, 561, 636  
     oxalate calculus, iii, 1008, 1025  
     phosphate calculus, iii, 1015  
     tetany and, v, 751  
 Calculus, biliary, iii, 573  
     bronchial, ii, 925  
     of pancreas, iii, 632  
     renal, iii, 1007  
     salivary, iii, 71  
     tartar, iii, 71  
     tuberculosis of kidney and, iii, 978  
     ureteral, iii, 1007  
 Calmette's immunization in rabies, i, 970  
 Cancer from x-rays, ii, 336  
 Cancerum oris, iii, 49  
 Capillaries in aortic insufficiency, iv, 223  
 Capillary bronchitis, ii, 957  
     thrombosis, iv, 537, 547  
 Capsular splenitis, iv, 943  
 Capsule, internal, lesion of, v, 276  
 Carbohydrates, absorption of, in intestines, iii, 290  
     diabetes mellitus and, ii, 677  
     fat formation and, ii, 603  
     metabolism of, ii, 596, 687  
     in diabetes mellitus, ii, 691  
 Carbon balance, ii, 560  
     bisulphide poisoning, ii, 392  
     dioxide in metabolism, ii, 618  
     metabolism of, ii, 560  
     moiety of protein, ii, 595, 622  
     monoxide poisoning, ii, 388  
 Carbuncles in diabetes mellitus, ii, 704  
 Carcinoma of adrenal glands, iv, 784  
 Carcinoma of ampulla of Vater, iii, 597  
     of appendix, iii, 386  
     of biliary ducts, iii, 593  
     of brain, v, 315  
     of choledochopancreatic duct, iii, 594  
     of colon, iii, 429  
     of duodenum, iii, 427  
     of gall-bladder, iii, 593  
     of ileum, iii, 428  
     of intestines, iii, 424  
     of jejunum, iii, 428  
     of kidney, iii, 1001  
     of lips, iii, 45  
     of liver, iii, 541  
     of lung, ii, 998  
     of lymph nodes, iv, 747  
     of lymphatic vessels, iv, 601  
     of mediastinum, ii, 1100  
     of mouth, iii, 52  
     of oesophagus, iii, 98  
     of pancreas, iii, 648  
     of pericardium, iv, 74  
     of pituitary body, iv, 804  
     of pleura, ii, 1061  
     of prostate, iii, 1038  
     secondary anemia and, iv, 659  
     of spinal cord, v, 196  
     of spleen, iv, 951  
     of stomach, iii, 199  
     of thoracic duct, iv, 588  
     of thymus, iv, 932  
     of thyroid gland, iv, 832  
     of tongue, iii, 66  
     in tuberculosis, i, 435  
 Cardiac arrhythmia, iv, 29, 286  
     psychical, iv, 287  
     in Stokes-Adams syndrome, iv, 305  
     in tuberculous meningitis, iv, 287  
 asthma, iv, 207  
 complications in lobar pneumonia, i, 255  
 crises in tabes dorsalis, v, 842  
 disease, congenital, iv, 323  
 endocarditis, iv, 162  
 epilepsy, v, 597  
 hypertrophy in myocarditis, iv, 121  
 insufficiency, iv, 191  
 nerves, paralysis of, v, 552  
 neuroses, iv, 296  
 nutrition, derangements of, iv, 108  
 syncope, iv, 88, 89  
 thrombosis, iv, 531, 536, 548  
 Cardialgia in hysteria, v, 683  
 Cardiolysis, iv, 66  
 Cardiopotosis, iii, 752  
 Cardiopulmonary murmurs, iv, 298  
 Cardiospasm, iii, 147  
     acute dilatation and, iii, 251  
     neurasthenia and, iii, 147  
 Cardiovascular disease, general considerations of, iv, 17  
     lesions in gout, ii, 745  
     system in lead poisoning, ii, 365  
     in rheumatic fever, i, 984  
 Caries, sicca, v, 189  
     spinal, v, 188

- Carious teeth, tuberculosis of cervical glands and, iv, 743
- Carotid artery, embolism of, iv, 567  
thrombosis of, iv, 567
- Carriers of bacillary dysentery, i, 771  
of diphtheria, i, 732  
of malarial fevers, ii, 99  
of scarlet fever, i, 862  
of typhoid fever, i, 76, 197
- Caseous pneumonia, i, 352, 424
- Casts in diabetes mellitus, ii, 704  
in fibrinous bronchitis, ii, 907, 909  
kidney and, iii, 774  
of pregnancy and, iii, 868  
in parenchymatous nephritis, iii, 863, 882, 901  
renal, iii, 798
- Catarrh, in chronic bronchitis, ii, 896  
postnasal, ii, 838  
suffocative, ii, 962
- Catarrhal appendicitis, iii, 372  
choolangitis, iii, 552, 559  
cholecystitis, iii, 562  
jaundice, iii, 448, 552  
laryngitis, ii, 868  
pharyngitis, ii, 844  
pneumonia, ii, 957  
pyelitis, iii, 953  
stomatitis, iii, 45  
ulcers, iii, 351
- Catatonie rigidity, v, 65
- Cavum Meckelii, tumors of, v, 340
- Cecum, rupture of peri-appendicular abscess into, iii, 365
- Cell embolism, iv, 561
- Cellulitis in scarlet fever, i, 878
- Centrum semiovale, lesion of, v, 275  
tumor in, v, 276, 334
- Cephaloceles, hydrocephalus and, v, 354
- Cephalomegalia, v, 988
- Cerebellar artery, posterior inferior, occlusion of, v, 288  
ataxia, v, 72  
Marie's hereditary, v, 130, 136  
ataxic gait, v, 74  
localization, v, 286  
syndrome, v, 286
- Cerebellum, agnesia of, v, 287  
atrophy of, v, 287  
lesion of, v, 284  
sclerosis of, v, 287  
tumors of, v, 338
- Cerebral abscess, v, 378  
empyema and, ii, 1045  
arteries, aneurism of, v, 398  
embolism of, v, 400  
rupture of, v, 400  
thrombosis of, v, 400  
arteriosclerosis, v, 393  
bloodvessels, diseases of, v, 387  
diplegia, v, 419  
disseminated sclerosis, v, 151  
hemiplegia, congenital, v, 409  
infantile, v, 412  
hemisphere, ganglia of, v, 278  
hemorrhage, v, 401, 410, 411, 414, 415  
in purpura, iv, 697
- Cerebral localization hypothesis of hysteria, v, 655  
nerves, diseases of, v, 471  
palsies, congenital, v, 410  
signs of Stokes-Adams syndrome, v, 307  
softening in apoplexy, v, 431  
symptoms of lead poisoning, ii, 367  
syphilis, diabetes insipidus and, ii, 722
- Cerebrospinal disseminated sclerosis, v, 154  
fever, i, 594  
hydrocephalus and, v, 357  
fluid, v, 351  
circulation of, v, 352  
examination of, v, 174  
lymphocytosis of, v, 177  
pressure of, v, 175  
in syphilis, v, 828  
meningitis, acute, v, 167  
epidemic, i, 594  
syphilis, v, 823  
vomiting, iii, 143
- Cervical adenitis, acute, i, 1012  
glands, tuberculosis of, iv, 743  
heart, iv, 346  
lymphadenitis, iv, 733, 734  
plexus, diseases of, v, 542  
ribs, v, 554
- Cervico-occipital neuralgia, v, 736
- Cestode infection, ii, 245
- Chalicosis, ii, 976
- Chancre, ii, 154
- Charbon, i, 639
- Charcot-Leyden crystals in asthma, ii, 939
- Charcot-Marie-Tooth type of progressive muscular atrophy, v, 109
- Cheinokinesesthetic agraphia, v, 75
- Chest, deformities of, hypertrophy of heart and, iv, 181  
skin of, in aneurism of aorta, iv, 495
- Cheyne-Stokes respiration, ii, 834  
in acute meningitis, v, 170  
in apoplexy, v, 436  
in chronic nephritis, iii, 905  
in fat embolism, iv, 560  
in Stokes-Adams syndrome, iv, 307
- Chicken mites, ii, 323
- Chickenpox, i, 849
- Chigger, ii, 324
- Chigoe, ii, 324
- Chloroma, iv, 679, 681  
of brain, v, 315
- Chlorosis, iv, 645  
cerebral thrombosis, and, v, 400  
exophthalmic goitre and, iv, 851  
gastric ulcer and, iii, 171  
hyperacidity and, iii, 114  
hyperthyroidism and, iv, 648  
hypoplasia of aorta and, iv, 432  
tropical, ii, 271
- Choked disk, v, 36  
in brain tumor, v, 326, 338  
in cerebral aneurism, v, 399  
stages of, v, 327
- Cholangitis, catarrhal, acute, iii, 552

- Cholangitis, catarrhal, cholelithiasis and,  
     iii, 570, 574, 583  
     chronic, iii, 559  
     suppurative, iii, 556
- Cholecystitis, abscess of liver and, iii, 497  
     acute, iii, 561  
     chronic, iii, 568  
     peritonitis and, iii, 714  
     in typhoid fever, i, 119
- Choledochopancreatic duct, carcinoma of,  
     iii, 594
- Cholelithiasis, iii, 569  
     appendicitis and, iii, 571  
     atony of stomach and, iii, 161  
     carcinoma of gall-bladder and, iii, 593  
     cholangitis and, iii, 570, 574, 583  
     hyperacidity and, iii, 114  
     metabolism and, iii, 571  
     pancreas and, iii, 613  
     pericholecystic adhesions and, iii, 583  
     pregnancy and, iii, 571
- Cholemia, iii, 454
- Choleperitoneum, iii, 670
- Cholera, Asiatic, i, 672  
     infantum, i, 776; iii, 341  
     morbus, iii, 331  
     nostras, iii, 331  
     vibrio, i, 673
- Cholesteatoma of brain, v, 316
- Cholesterin cysts of spleen, iv, 952
- Choluria, iii, 828
- Chondritis, gonococcal, i, 760
- Chondrodystrophia fetalis hypoplastica, v,  
     958, 959
- Chondrosarcoma of lymphatic vessels, iv,  
     601
- Chordæ tendineæ, anomalous, iv, 354
- Chordoma of brain, v, 317
- Chorea, acute, v, 774  
     chronic, v, 787  
     degenerative, v, 787  
     electric, v, 787  
     habit, v, 784  
     Huntington's, v, 787  
     insaniens, v, 779  
     minor, v, 774  
     mollis, v, 780  
     nephritis and, iii, 850  
     pericarditis and, iv, 42  
     posthemiplegic, v, 466  
     rheumatic fever and, i, 993; v, 775  
     Sydenham's, v, 774
- Choroidal tubercles in acute miliary tuber-  
     culosis, i, 419
- Choroidoretinitis, syphilitic, v, 475
- Chromaffin bodies, iv, 778  
     system, diabetes mellitus and, ii, 682  
     tissue, iv, 777
- Chvostek's phenomenon v, 71  
     in myxœdema, iv, 893  
     in sporadic cretinism, iv, 885  
     in tetany, v, 758
- Chyle angiomas, iv, 586  
     cysts of thoracic duct, iv, 586  
     stone in thoracic duct, iv, 582
- Chyliform ascites, iii, 661  
     pleural fluids, ii, 1057
- Chylopericardium, iv, 590
- Chylothorax, ii, 1057; iv, 590  
     in anomalies of thoracic duct, iv, 580  
     in carcinoma of thoracic duct, iv, 589  
     in sarcoma of thoracic duct, iv, 588  
     in thrombosis of thoracic duct, iv, 581  
     traumatic, ii, 1059
- Chylous ascites, iii, 660; iv, 589  
     pleural fluids, ii, 1057
- Chyluria, iii, 805; iv, 591  
     in carcinoma of thoracic duct, iv, 589  
     in filariasis, ii, 309  
     in lymphangitis, iv, 595  
     in thrombosis of thoracic duct, iv, 581
- Cicatricial stenosis of pylorus, iii, 262
- Cimex lectularius, ii, 327
- Circulatory changes in pituitary body, iv,  
     804  
     disturbances of adrenal glands, iv, 782  
     of kidney, iii, 787  
     of lungs, ii, 945  
     neurasthenia and, v, 631  
     of thymus, iv, 909  
     organs, syphilis of, ii, 180  
     system in arthritis deformans, v, 914  
     in chlorosis, iv, 650  
     gastric ulcer and, iii, 171  
     in influenza, i, 544  
     in pernicious anemia, iv, 632  
     in plague, i, 628  
     in rickets, ii, 790  
     in tuberculosis, i, 359, 384  
     in typhoid fever, i, 81, 124, 131  
     in typhus fever, i, 926, 930  
     in uremia, iii, 838
- Circumflex nerve, paralysis of, v, 546
- Circumscribed pericarditis, iv, 42  
     saccular aneurism, iv, 472
- Circumspection, v, 43
- Cirrhosis of liver, iii, 505  
     arthracotic, iii, 531  
     cardiac, iii, 483  
     malarial, iii, 530  
     of lungs, ii, 969  
     of stomach, iii, 273
- Cirroid aneurism, iv, 437, 486
- Cladotrix buccalis in mouth, iii, 40
- Clergyman's sore throat, ii, 844
- Clonorchiasis, Asiatic, ii, 230
- Clonorchis, ii, 229
- Clubbed fingers, v, 981
- Coarctation of aorta, iv, 420
- Cocaine habit, ii, 445
- Coccygeal plexus, neuralgia of, v, 741
- Cochlear nerve, diseases of, v, 506
- Coin sound in pneumothorax, ii, 1081
- Colic, appendicular, iii, 373  
     biliary, iii, 576  
     gall-stone, iii, 576  
     intestinal, iii, 303  
     lead, iii, 306  
     mucous, iii, 437
- Colitis, iii, 336  
     amœbic, ii, 17  
     in lobar pneumonia, i, 254  
     membranous, iii, 437  
     mucous, iii, 33, 437, 757



- Colitis, simple, iii, 336  
 Colles' law in syphilis, ii, 153  
 Colloid goitre, iv, 839  
   matter of thyroid gland, iv, 823  
 Colodysentery, i, 775  
 Colon bacillus infections, i, 199  
   carcinoma of, iii, 429  
   congenital dilatation of, iii, 321  
   disease of, peritonitis and, iii, 673  
   displaced, iii, 752  
   distension of, kidney infections and, iii, 952  
 Color blindness, v, 25  
 Colorless stools, iii, 294  
 Combustion products poisoning, ii, 388  
 Compensatory emphysema of lungs, ii, 982  
   hypertrophy of heart, iv, 176  
 Complement fixation, i, 57  
   test in glanders, i, 654  
   in gonococcus infections, i, 744  
 Compression of intestines, iii, 394  
   myelitis, v, 229  
   paralysis in syphilis, ii, 162  
 Compomyia marcellaria, ii, 328  
 Condyloma, syphilitic, ii, 165  
 Confusion, v, 43  
 Congenital anomalies of liver, iii, 455  
   aortic atresia, iv, 398, 399  
   insufficiency, iv, 407  
   stenosis, iv, 398, 399  
   cardiac disease, iv, 323  
     convulsions and, v, 794  
   cerebral hemiplegia, v, 409  
   chronic interstitial nephritis, iii, 891  
   cyanosis, iv, 333  
   cystic kidney, iii, 786  
   cysts of brain, v, 318  
   diverticula of intestines, iii, 421  
   elephantiasis, iv, 597  
   hypertrophic stenosis of pylorus, iii, 269  
   hypertrophy of heart, iv, 351  
   malformations of œsophagus, iii, 78  
   mitral atresia, iv, 406  
   obliteration of biliary ducts, iii, 547  
   osteosclerosis, v, 968  
   pulmonary atresia, iv, 388  
   pylorospasm, iii, 269  
   rhabdomyoma, iv, 351  
   rickets, ii, 795; v, 955  
   stenosis of intestine, iii, 408  
     of œsophagus, iii, 96  
   syphilis, ii, 187  
     of liver, ii, 177  
   tricuspid atresia, iv, 403  
   insufficiency, iv, 470  
   stenosis, iv, 403  
 Congestion of bowels, constipation and, iii, 312  
   of liver, iii, 478  
   of lungs, ii, 945  
   of spleen, iv, 940  
   of thymus, iv, 909, 912  
   of thyroid gland, iv, 825  
 Conjugate deviation in apoplexy, v, 438  
 Conjunctivitis in angioneurotic œdema, iv, 1007  
   in cerebrospinal meningitis, i, 604  
   gonorrhœal, i, 746  
   in lobar pneumonia, i, 262  
   in smallpox, i, 812  
 Conorhinus sanguisuga, ii, 327  
 Consciousness, v, 41, 42, 43  
 Constipation, iii, 309  
   atonic, iii, 313  
   spastic, iii, 313  
 Constriction of intestines, iii, 394  
 Continuous gastromyorrhœa, iii, 131  
 Contraction, premature, of heart, iv, 84, 91, 95, 100  
 Contractures in amyotrophic lateral sclerosis, v, 99  
   in facial paralysis, v, 502  
   in hemiplegia, v, 464  
   in hereditary spastic spinal paralysis, v, 106  
   in hysteria, v, 651  
   of muscles, v, 68  
   in progressive neural muscular atrophy, v, 110  
   in traumatic neuroses, v, 151  
 Conus stenosis, left-sided, iv, 400  
 Convulsions, central, lesions of, v, 274  
 Convulsions in abscess of brain, v, 380, 381  
   in acute meningitis, v, 170  
   miliary tuberculosis, i, 421  
   nephritis, iii, 394  
   in Addison's disease, iv, 793  
   in anemia of brain, v, 392  
   in arteriosclerosis, iv, 462  
     of brain, v, 397  
   in brain tumor, v, 329, 333  
   in cardiac insufficiency, iv, 209  
   in cerebrospinal meningitis, i, 605  
   in chronic interstitial nephritis, iii, 908  
   in fat embolism, iv, 560  
   in general paresis, v, 857  
   hemiplegia and, v, 415  
   in hysteria, v, 673  
   infantile, v, 790  
   in lead poisoning, ii, 368  
   in lesion of pons, v, 290  
   in lobar pneumonia, i, 223  
   in measles, i, 907  
   in mercury poisoning, ii, 380  
   in pachymeningitis interna, v, 165  
   postnatal infantile hemiplegia and, v, 415  
   in rabies, i, 965  
   in typhoid fever, i, 138  
   in uremia, iii, 835  
 Convulsive tic, v, 784  
 Coordination, disturbances of, v, 71  
 Copper poisoning, ii, 386  
 Cor biatriatum triloculare, iv, 372  
   biloculare, iv, 370  
   biventriculare triloculare, iv, 374  
 Coronary arteries, accessory, iv, 439  
   anomalies of, iv, 437  
   in arteriosclerosis, iv, 463  
   atheroma of, cardiac insufficiency and, iv, 195

- Coronary arteries, cirroid aneurism of, iv, 437  
 embolism of, iv, 195, 569  
 sclerosis of, myocarditis and, iv, 109  
 thrombosis of, iv, 109, 195, 569  
 disease, angina pectoris and, iv, 289  
 spasm, angina pectoris and, iv, 293  
 Corpora quadrigemina, tumors of, v, 336  
 striata, lesion of, v, 278  
 Corpus callosum, disease of, v, 276  
 tumor of, v, 276, 355  
 Corrigan pulse, iv, 220, 224  
 Corrosive œsophagitis, iii, 81  
 Cortex of brain, v, 265, 273  
 Cortical blindness, v, 54  
 deafness, v, 33  
 Coryza, acute bronchitis and, ii, 881  
 in measles, i, 906  
 in migraine, v, 718  
 in tuberculosis, i, 506  
 Coughing, mechanism of, ii, 828  
 Courvoisier's law, iii, 579  
 Cowpox, i, 822  
 Cramps in occupation neuroses, v, 744  
 professional, v, 741  
 Cranial base, tumors of, v, 340  
 nerves in tabes dorsalis, v, 841  
 Cranium in rickets, ii, 785  
 Creatin in fasting, ii, 614  
 in protein metabolism, ii, 589  
 Creatinin, excretion of, iii, 813  
 Cremaster reflex, anomalies of, v, 78  
 Cretinism, iv, 881  
 Crisis in lobar pneumonia, i, 240  
 Croupous pneumonia, i, 202  
 Crura cerebri, lesion of, v, 281  
 tumors of, v, 335  
 Crural neuralgia, v, 740  
 Cryoscopy in lobar pneumonia, i, 233  
 of urine, iii, 792  
 Cryptogenic anemia, iv, 619  
 Crystallization of renal calculi, iii, 1014  
 Crystals in urine, iii, 800  
 Crytometry in tuberculosis, i, 404  
 Ctenocephalus canis, ii, 325  
 Curschmann's spirals in asthma, ii, 939  
 Cutaneous actinomycosis, i, 1040  
 burns, duodenal ulcer and, iii, 349  
 diphtheria, i, 699  
 lesions in syphilis, ii, 163, 168  
 in uremia, iii, 840  
 reaction in syphilis, ii, 193  
 reflexes, anomalies of, v, 77  
 syphilides, ii, 150, 184  
 Cyanosis, congenital, iv, 333  
 Cylindrical aneurism, iv, 479  
 Cylindroids in urine, iii, 800  
 Cystic distension of appendix, iii, 386  
 duct, cholelithiasis and, iii, 580  
 obstruction of, iii, 580  
 kidney, congenital, iii, 786  
 lymphangiomata, iv, 586  
 pyelitis, iii, 354  
 softening of thymus, iv, 912  
 Cysticercosis, ii, 263  
 Cysticercus bovis, ii, 247, 264  
 cellulose, ii, 249, 263  
 cyst of peritoneum, iii, 736  
 of spleen, iv, 954  
 fasciolaris, ii, 252  
 tenuicollis, ii, 264  
 Cystin calculus, iii, 1009  
 Cystinuria, ii, 533, 656; iii, 817  
 Cystitis in appendicitis, iii, 371  
 gonococcal, i, 762  
 kidney infections and, iii, 952  
 in smallpox, i, 812  
 tuberculosis of kidney and, iii, 978  
 in typhoid fever, i, 143  
 Cystoglioma of brain, v, 313  
 Cystosarcoma of brain, v, 315  
 Cystoscopy, iii, 938  
 in hypernephroma, iii, 998  
 Cysts of brain, v, 318  
 chyle, of thoracic duct, iv, 586  
 of liver, iii, 547  
 of lymph nodes, iv, 586, 753  
 of mediastinum, ii, 1101  
 of mouth, iii, 52  
 necrosis of pancreas and, iii, 619  
 ovarian, acute peritonitis and, iii, 673  
 of peritoneum, iii, 733-739  
 of spleen, iv, 951  
 of thymus, iv, 931, 933  
 Cytodiagnosis, ii, 1030  
 Cytological examination of cerebrospinal fluid, v, 176  
 Cytology of pleural fluids, ii, 1030  
 in tuberculosis, i, 422  
 Cytolysis, auto-intoxication and, ii, 504  
 Cytoryctes vaccinae, i, 825  
 variolae, i, 786

## D

- DACRYOADENITIS, gonococcal, i, 764  
 Davainea asiatica, ii, 246, 255  
 madagascariensis, ii, 246, 255  
 Day blindness, v, 480  
 Deafness, mind, v, 54  
 word, v, 54  
 Decubital ulcers, iii, 351  
 of œsophagus, iii, 84  
 Degeneration, fatty, ii, 604  
 of liver, iii, 466  
 of lymph nodes, iv, 746  
 of myocardium, iv, 108  
 of pituitary body, iv, 804  
 progressive lenticular, v, 278  
 reaction of, v, 72  
 Degenerative myelitis, v, 236  
 Deglutition pneumonia, ii, 958  
 Delirium cordis, iv, 106  
 tremens, ii, 408  
 in uremia, iii, 837  
 Delusions, v, 56  
 Demacentor andersoni, ii, 317, 318  
 Dementia, anomalies of conduct in, v, 67  
 in arteriosclerosis, iv, 462  
 paralytica, v, 850

- Dementia præcox, hysteria and, v, 694,  
     695  
 Demodex folliculorum, ii, 321  
 Dengue, i, 950  
 Depression, pathological, v, 60  
 Dercum's disease, v, 817  
 Dermanyssus, ii, 323  
 Dermatitis, bullous, in hysteria, v, 686  
     coccidioides, i, 1050  
     in dermatomyositis, v, 877  
     from x-rays, ii, 335  
 Dermatocentor andersonii, i, 1021  
 Dermatomyositis, v, 875  
 Dermatoses of mouth, iii, 53  
 Dermoid cyst of mediastinum, ii, 1101  
     of mouth, iii, 53  
     of peritoneum, iii, 739  
     of spleen, iv, 953  
     of thymus, iv, 933  
     tumors of brain, v, 316  
 Dextrocardia, iv, 347  
 Diabetes, bronze, ii, 681  
     insipidus, ii, 721  
         calculus of pancreas and, iii, 635  
         carcinoma of pancreas and, iii,  
             650, 652  
     lipogenous, ii, 676  
     mellitus, ii, 674  
         apoplexy and, v, 427  
         cyst of pancreas and, iii, 645  
         diabetes insipidus and, ii, 724  
         gangrene of lung and, ii, 991  
         lobar pneumonia and, i, 207  
         pancreas and, iii, 604, 619, 630  
         tuberculosis and, i, 320, 435  
 Diabetogenous obesity, ii, 676  
 Diacetic acid in diabetes mellitus, ii, 702  
 Diadokokinesis, v, 72  
 Diamino-acids, ii, 656  
 Diaminuria, ii, 656  
 Diaphragm, immobility of, in adherent  
     pericardium, iv, 61  
     perforation of, in purulent pleuritis, ii,  
         1045  
 Diaphragmatic neuralgia, v, 737  
     pleuritis, ii, 1049  
 Diarrhoea, iii, 322  
     in childhood, iii, 337, 340, 341  
 Diastole, nature of, iv, 22, 32  
 Diastolic collapse of veins, iv, 62  
 Diazo reaction in typhoid fever, i, 141  
 Dibothriocephalus cordatus, ii, 246, 257  
     latus, ii, 245, 255  
 Dichromates, v, 25  
 Dicrocoelium, ii, 229, 234  
 Dicrotism, iv, 35  
     in typhoid fever, i, 124  
 Dietl's crises in nephroptosis, iii, 756, 759  
 Digestive gastrosuccorrhœa, iii, 128  
     tract in lead poisoning, ii, 364  
     tuberculosis of, i, 360  
 Dilatation aneurism, iv, 472, 482  
     of aorta, dynamic, iv, 513  
     in diseases of valves of heart, iv, 214  
     of heart, iv, 30, 189  
         defects of interauricular septum  
         and, iv, 359  
 Dilatation of hepatic artery, iii, 158  
     veins, iii, 459  
 idiopathic, iii, 320  
 of œsophagus, iii, 85, 90, 91  
 of pulmonary artery, iv, 398  
     defects of interauricular sep-  
     tum and, iv, 359  
 of stomach in lobar pneumonia, i, 254  
     in tuberculosis, i, 502  
 tricuspid insufficiency and, iv, 247  
 Diphtheria, i, 689  
     antitoxin in, i, 54, 715  
     apoplexy and, v, 417  
     arteriosclerosis and, iv, 454  
     bronchopneumonia and, ii, 958  
     cardiac insufficiency and, iv, 196  
     carriers, i, 732  
     cutaneous, i, 699  
     enlargement of thymus and, iv, 915  
     of eye, i, 699  
     laryngeal, i, 698  
     of lungs, i, 700  
     multiple neuritis and, v, 575  
     myocarditis and, iv, 108, 116  
     nasal, i, 698  
     nephritis and, iii, 849  
     pharyngeal, i, 696  
     tuberculosis and, i, 321, 436  
     of uterus, i, 699  
 Diphtheritic gastritis, iii, 236  
     pyelitis, iii, 954  
 Diplegia, cerebral, v, 419  
 Diplococcus catarrhalis, i, 595  
     intracellularis meningitidis, i, 594  
     rheumaticus, i, 975  
 Diplogonoporus, ii, 246, 258  
 Diplopia in disseminated sclerosis, v, 150  
     in interstitial nephritis, iii, 910  
     monocular, v, 480  
     in myasthenia gravis, v, 802  
 Dipsomania, ii, 423  
 Dipterous infection, ii, 328  
 Dipylidium caninum, ii, 246, 255  
 Dirofilaria, ii, 314  
 Dirt eating, ii, 271  
 Dislocated spleen, iv, 938  
 Disorientation, v, 44  
 Dissecting aneurism, iv, 472, 487, 489  
 Disseminated gummatous sporotrichosis, i,  
     1056  
     myelitis, v, 235  
     sclerosis, v, 140  
 Dissociation, v, 51  
 Distension in intestinal indigestion, iii, 301  
     of stomach, cardiac arrhythmia and, iv,  
         287  
 Distoma hepaticum, ii, 235  
 Distomatosis, ii, 225  
     hepatic, ii, 229  
     intestinal, ii, 237  
     ophthalmic, ii, 235  
     pulmonary, ii, 226  
     renal, ii, 239  
 Distomiasis, ii, 225  
 Diver's paralysis, ii, 345  
 Diverticula of intestines, iii, 420  
     of œsophagus, iii, 92

- Diverticulitis, iii, 423  
 Diverticulum of heart, iv, 351  
   of pericardium, iv, 346  
   of Vater, biliary infections through, iii, 550  
 Dracontiasis, ii, 303  
 Dracunculosis, ii, 303  
 Dream states, v, 689  
 Dropsy, renal, ascites and, iii, 659  
 Drunkenness, pathological, ii, 424  
 Dry pleurisy, ii, 1051  
 Duchenne-Erb palsy, v, 551  
 Ductless glands, diseases of, iv, 777  
 Ductus arteriosus Botalli, absence of, iv, 420  
   aneurism of, iv, 419  
   anomalies of, iv, 409, 420  
   endarteritis and, iv, 413  
   left subclavian artery from, iv, 436  
   patency of, iv, 409  
 Dum-dum fever, iv, 965  
 Duodenal ulcer, iii, 191, 344  
   chronic peritonitis and, iii, 714  
   intestinal hemorrhage and, iii, 307  
   subphrenic abscess and, iii, 701  
   pyopneumothorax and, iii, 710  
 Duodenitis, necrosis of pancreas and, iii, 612  
 Duodenum, carcinoma of, iii, 427  
   perforation of, in necrosis of pancreas, iii, 619  
   stenosis of, iii, 262  
 Dura mater, diseases of, v, 163  
 Dwarf tapeworm, ii, 252  
 Dysacusis, v, 33  
 Dysarthria, v, 74  
 Dyschiria, v, 268  
 Dyschondroplasia, v, 969  
 Dyschromatopsia in brain tumor, v, 329  
 Dysentery, amœbic, ii, 17, 33, 34  
   bacillary, i, 766  
 Dysesthesia, v, 32  
   in brain tumor, v, 339  
 Dyspepsia, nervous, iii, 164  
 Dyspeptic cirrhosis of liver, iii, 509  
 Dysphonia, spastic, v, 522  
 Dysplastic periosteal achondroplasia, v, 969  
 Dyspnœa, bulbar, iv, 40  
   nervous, iv, 37  
   reflex, iv, 38  
   toxic, iv, 37  
 Dyspragia, angiosclerotic, iii, 437  
 Dyspraxia, v, 75  
 Dysthymia, v, 60  
 Dystrophy, progressive muscular, v, 117  
   facio-scapulo-humeral, v, 122  
   hereditary, v, 123  
   infantile, v, 123  
   pseudo-hypertrophic, v, 122  
   of Zimmerlin, v, 123
- E**
- EAR in acromegaly, iv, 811  
   in cerebrospinal meningitis, i, 604, 609  
   in measles, i, 905  
   in myxœdema, iv, 891  
   in tuberculosis, i, 431  
   in typhoid fever, i, 140  
 Echinocynchus, ii, 297  
 Echinococcosis, ii, 264  
 Echinococcus cyst of brain, v, 319  
   of mediastinum, ii, 1109  
   disease, ii, 264  
   of pleura, ii, 1065  
   granulosus, ii, 263  
   kidney infections and, iii, 952  
   multilocularis, ii, 266  
   of myocardium, iv, 147  
   of thyroid gland, iv, 831  
 Echinostoma ilocanum, ii, 238  
 Echolalia, v, 65  
 Echopraxia, v, 65  
 Eclampsia in kidney of pregnancy, iii, 869  
   parathyroid gland and, iv, 903  
 Ectasia of thoracic duct, iv, 585  
 Ecthyma in congenital syphilis, ii, 190  
 Ectopia cordis, iv, 346  
 Eczema in gout, ii, 748  
   of lips, iii, 45  
   of mouth, iii, 54  
   in vaccination, i, 843  
 Eelworm infection, ii, 287  
 Egyptian chlorosis, ii, 271  
   hematuria, ii, 239  
 Electric chorea, v, 787  
 Electrical hyperexcitability in tetany, v, 757  
   reactions in diseases of peripheral nerves, v, 535  
   in multiple neuritis, v, 582  
   in progressive spinal muscular atrophy, v, 85  
 Electricity, ii, 339  
 Electrocardiograph, iv, 80  
   in congenital disease of heart, iv, 444  
 Elephantiasis Arabum, iv, 596  
   in filariasis, ii, 310  
   lymphangiectatica, iv, 596-598  
   neuromatosa, v, 571  
   in thrombosis of thoracic duct, iv, 581  
 Elephantoid fever, ii, 308  
 Emboli, endogenous, iv, 553  
   exogenous, iv, 553  
   infective, iv, 557  
 Embolic aneurisms, iv, 557  
   ulcers, iii, 350  
 Embolism, iv, 552  
   air, iv, 559  
   aneurism and, iv, 476  
   of aorta, iv, 565  
   in aortic insufficiency, iv, 228  
   apoplexy and, v, 418, 422, 424, 427, 428, 431  
   of carotid artery, iv, 567  
   cell, iv, 561  
   of cerebral arteries, v, 400  
   of coronary arteries, iv, 569

- Embolism, crossed, iv, 553  
 fat, iv, 560  
 gastric ulcer and, iii, 171  
 of hepatic artery, iii, 458; iv, 569  
 infarcts and, iv, 556, 946  
 infective, iv, 557  
 in lobar pneumonia, i, 258  
 mercurial, iv, 561  
 mesenteric, iii, 307, 434; iv, 565  
 in mitral insufficiency, iv, 236  
   stenosis, iv, 242  
 paradoxical, iv, 363, 553  
 paraffin, iv, 562  
 parasitic, iv, 558  
 of peripheral arteries, iv, 568  
 of portal vein, iii, 463  
 postnatal infantile hemiplegia and, v, 418  
 of pulmonary artery, iv, 562  
 in pyemia, i, 565  
 of renal artery, iv, 564  
 of retinal arteries, iv, 570  
 retrograde, iv, 553  
 of splenic artery, iv, 564  
 of subclavian artery, iv, 567  
 in tuberculosis, i, 433  
 tumor cells and, iv, 557  
 in typhoid fever, i, 129  
 venous, iv, 552
- Embryocardia, iv, 279
- Embryonic tumors of kidney, iii, 1003
- Emmetropia, v, 36
- Emotional states, anomalies of, v, 59
- Emphysema, cardiac insufficiency and, iv, 199  
 in diphtheria, i, 693  
 of lungs, ii, 979  
 of mediastinum, ii, 1110  
 in tuberculosis, i, 430  
 in whooping cough, i, 660
- Emphysematous liver, iii, 466
- Empyema, ii, 1040  
 abscess of brain and, v, 379  
 chronic, ii, 1052  
 encysted, ii, 1049  
 of gall-bladder, iii, 563, 566  
 in influenza, i, 542  
 interlobar, ii, 1050  
 in lobar pneumonia, i, 248  
 necessitatis, ii, 1044  
 pulsating, ii, 1043  
 of renal pelvis, iii, 937  
 subphrenic abscess and, iii, 701  
 in tuberculosis, i, 431  
 tuberculous, ii, 1048  
 in typhoid fever, i, 123
- Encephalic tuberculoma of brain, v, 319
- Encephalitis, v, 359  
 acute, v, 360, 361  
   hemorrhagic, v, 367  
   hyperplastic, v, 370  
   non-suppurative, v, 370  
 congenita, v, 361, 370  
 epilepsy and, v, 599  
 experimental, v, 371  
 in infancy and childhood, v, 376  
 influenzal, i, 543; v, 362
- Encephalitis, neonatorum, v, 370  
 postnatal infantile hemiplegia and, v, 416  
 progressive, v, 361  
 pyogenic, v, 363  
 subacute, v, 360, 361  
 superior, v, 490
- Encephalomyelitis, acute, v, 369
- Encephalopathy in lead poisoning, ii, 367
- Enchondroma of brain, v, 317  
 of lung, ii, 997
- Encysted empyema, ii, 1049  
 pleuritis, ii, 1049  
 tuberculosis of peritoneum, iii, 723
- Endarteritis, acute gummatous, ii, 181  
 pulmonary, iv, 413  
 in cerebral syphilis, v, 824  
 deformans, iv, 452
- ductus arteriosus Botalli and, iv, 413  
 obliterative, ii, 181; iv, 458
- Endocarditis, acute, iv, 148  
 aortic insufficiency and, iv, 218  
   stenosis and, iv, 229  
 in arthritis deformans, v, 914  
 chorea and, v, 776, 781  
 chronic, iv, 212  
   septic, iv, 148  
 coarctation of aorta and, iv, 429  
 coronary embolism and, iv, 569  
 in erysipelas, i, 587  
 gonorrhoeal, i, 750  
 in influenza, i, 544  
 in lobar pneumonia, i, 255  
 in lymphoid leukemia, iv, 675  
 malignant, iv, 161-163  
 in measles, i, 907  
 mesenteric embolism and, iii, 435  
 mitral insufficiency and, iv, 232  
   stenosis and, iv, 239  
 necrotic, iv, 151  
 primary, iv, 153  
 pulmonary stenosis and, iv, 258  
 in purpura, iv, 706  
 recurrent, iv, 148  
 in rheumatic fever, i, 985  
 in scarlet fever, i, 877  
 simple, iv, 148, 158  
 subaortic stenosis and, iv, 399  
 syphilitic, ii, 183  
 tricuspid insufficiency and, iv, 244  
 in tuberculosis, i, 433  
 in typhoid fever, i, 126  
 ulcerative, iv, 148, 151  
 vegetative, iv, 151  
 verrucose, iv, 150
- Endogenous emboli, iv, 553
- Endolymphangitis, iv, 595
- Endophlebitis of portal vein, iii, 460, 465
- Endothelioma of brain, v, 313  
 of lymph nodes, iv, 747  
 lymphangiomatous, iv, 600  
 of lymphatic vessels, iv, 600, 601  
 of mediastinum, ii, 1100  
 of thoracic duct, iv, 588
- Endotoxin, i, 33
- Energy metabolism, ii, 564  
 muscular, ii, 621, 631

- Enteralgia, iii, 305  
   in hysteria, v, 683  
 Enteritis, iii, 328, 334  
   lymphadenitis and, iv, 736  
 Enterocolitis, diarrhoea and, iii, 324  
 Enterodysentery, i, 775  
 Enterokinase, iii, 290  
 Enteroptosis, iii, 747  
   in atony of stomach, iii, 162  
 Enterostenosis, iii, 753  
 Entomœbæ Blattæ, ii, 22  
   coli, ii, 22, 23, 36, 45  
   hartmanni, ii, 19  
   histolytica, ii, 22, 23, 26, 46  
   hominis, ii, 22  
   tetragena, ii, 24, 47  
 Enzymes, defensive, ii, 651  
   intracellular, ii, 663  
   inverting, iii, 290  
   protective, ii, 651  
   spleen and, iv, 934  
 Eosinophiles in myeloid leukemia, iv, 668  
 Eosinophilia, iv, 615  
 Ependymal glioma of brain, v, 311  
   inflammation, hydrocephalus and, v, 358  
 Epieritic sensibility, v, 17  
 Epidemic cerebrospinal meningitis, i, 594  
 Epididymis, tuberculosis of, i, 366  
 Epigastric reflexes, v, 78  
 Epilepsy, v, 592  
   alcoholism and, ii, 408; v, 593, 596  
   apoplexy and, v, 421  
   in brain tumor, v, 331, 333  
   cerebral diplegia and, v, 421  
   in disseminated sclerosis, v, 153  
   idiopathic, v, 592  
   infantile convulsions and, v, 594  
   Jacksonian, v, 592  
   migraine and, v, 615, 711  
   parathyroid glands and, iv, 903  
   petit mal, v, 604  
   in pseudo-sclerosis, v, 159  
   in Raynaud's disease, iv, 988  
 Epileptic insanity, traumatism and, v, 647  
   psyche, v, 598, 612  
 Epileptoid attacks in lesion of centrum semiovale, v, 275  
 Epinephritis, iii, 970  
 Epiphyseal cartilages in achondroplasia, v, 958  
 Epiploitis, iii, 695  
 Epithelial casts, iii, 799  
   cells in tuberculosis of kidney, iii, 984  
 Epithelioid tuberculosis of lymph nodes, iv, 741  
 Epithelioma of brain, v, 315  
 Epitrochlear gland, tuberculosis of, iv, 744  
 Equilibrium, sense of, v, 18  
 Erb's phenomenon, v, 71  
   sign in tetany, v, 757  
   syphilitic paraplegia, v, 246, 828  
 Erepsin, iii, 290  
 Ergotismus, ii, 468  
 Erosion aneurism, iv, 472  
 Eructation, nervous, iii, 141  
 Erysipelas, i, 585  
 Erysipelas, lymphangitis and, iv, 592  
   of mouth, iii, 54  
   in myeloid leukemia, iv, 670  
   nephritis and, iii, 850  
   typhoid fever and, i, 158  
 Erythema, acute endocarditis and, iv, 155  
   in arsenic poisoning, ii, 375  
   in congenital syphilis, ii, 189  
   gonococcal, i, 764  
   in Henoch's purpura, iv, 709  
   infectiosum, i, 916  
   nodosum, gonococcal, i, 764  
   nephritis and, iii, 850  
   in rheumatic fever, i, 997  
   in pellagra, ii, 482  
   purpura and, iv, 705  
   rheumatica and, iv, 703  
   in rheumatic fever, i, 996  
   in smallpox, i, 799  
   in typhoid fever, i, 100  
 Erythematous scleroderma, iv, 1017  
 Erythrocytosis megalosplenica, iv, 682  
 Erythromelalgia, iv, 1025  
   in arteriosclerosis, iv, 465  
 Erythrospia, v, 480  
 Estivo-autumnal parasite, ii, 56, 59  
 Eustachian valve, iv, 327  
 Eustrongylus gigas, ii, 316  
   kidney infections and, iii, 952  
 Exaltation, pathological, v, 60  
 Excitability, affective, pathological, v, 62  
 Exogenous emboli, iv, 553  
 Exophthalmic goitre, iv, 849  
 Exophthalmos in congenital cyanosis, iv, 339  
   in exophthalmic goitre, iv, 862  
   in hypopituitarism, iv, 817  
   in ocular paralysis, v, 485  
 Extracerebellar tumors, v, 338  
 Extradural hemorrhage, v, 257  
 Extraperitoneal subphrenic abscess, iii, 705  
 Extrasystole, iv, 84  
 Exudate in ascites, iii, 659  
   in pericarditis, iv, 43  
 Eye lesions from electricity, ii, 342  
   from x-rays, ii, 336  
 Eyes in acromegaly, iv, 810  
   in aneurism of arch of aorta, iv, 494  
   diphtheria of, i, 699  
   in exophthalmic goitre, iv, 862  
   in hysteria, v, 684  
   in influenza, i, 545  
   in lymphoid leukemia, iv, 672  
   in measles, i, 905  
   in meningitis, cerebrospinal, i, 604, 609  
   posterior basic, i, 617  
   in myeloid leukemia, iv, 666  
   in myxœdema, iv, 893  
   in Raynaud's disease, iv, 987  
   in scarlet fever, i, 875  
   in typhoid fever, i, 139  
 Eye-grounds in abscess of brain, v, 380  
   in tumor of brain, v, 326

## F

- Facial atrophy in migraine**, v, 719  
   hemiatrophy, v, 991  
   irritability in rickets, ii, 789  
   nerve, paralysis of, v, 495  
     in tabes dorsalis, v, 841  
   neuralgia, v, 733  
   paralysis, v, 495  
     in hysteria, v, 679  
     otitis and, v, 497  
     poliomyelitis and, v, 497  
     polyneuritis and, v, 497  
   spasm, v, 504  
**Facies in acute peritonitis**, iii, 679  
   cholera, i, 683  
**Facio-scapulo-humeral progressive muscular dystrophy**, v, 122  
**Faget's sign in yellow fever**, i, 944  
**Falling sickness**, v, 592  
**Fallopian tubes**, abscess of, subphrenic  
   abscess and, iii, 701  
   tuberculosis of, i, 368  
**Family periodic paralysis**, v, 809  
**Farcy**, i, 649, 653  
**Fasciæ, tuberculosis of**, i, 373  
**Fasciola**, ii, 229, 235  
**Fasciolopsis**, ii, 237, 238  
**Fat, carbohydrates and**, ii, 603  
   embolism, iv, 560  
     thoracic duct and, iv, 582  
   metabolism, ii, 601  
     in obesity, ii, 605  
   necrosis, iii, 606  
   tapeworm, ii, 247  
**Fatigue, sense of**, v, 19, 32  
**Fatty ascites**, iii, 661  
   casts, iii, 800  
   degeneration of lymph nodes, iv, 746  
     in myocarditis, iv, 115  
     in pernicious anemia, iv, 625  
   feces, iii, 294  
   heart in myocarditis, iv, 115  
   infiltration of lymph nodes, iv, 746  
   liver, iii, 466  
   stools, iii, 294  
**Febricula**, i, 1009  
**Fecal concretions in appendicitis**, iii, 360  
   masses, intestinal obstruction from,  
     iii, 403  
**Feces**, iii, 292  
   acholic, iii, 294  
   colorless, iii, 294  
   fatty, iii, 294  
**Femoral glands, lymphadenitis of**, iv, 736  
   neuralgia, v, 740  
**Ferments in carcinoma of stomach**, iii, 211  
**Fetal passages in transposition of arterial trunks**, iv, 385  
   rickets, v, 955  
**Fetid bronchitis**, ii, 911  
**Fibrillary twitchings**, v, 70  
**Fibrillation, auricular**, iv, 86, 92, 96, 99  
   of heart, insufficiency and, iv, 192  
**Fibrinous bronchitis**, ii, 907  
   pleuritis, ii, 1005  
   pyelitis, iii, 954  
**Fibroid degeneration of myocardium**, iv,  
   114  
   nodules, endocarditis and, iv, 155  
   tumors of pericardium, iv, 76  
**Fibroma of lung**, ii, 998  
   of peritoneum, iii, 731  
   of spleen, iv, 950  
   of tongue, iii, 66  
**Fibromyoma of peritoneum**, iii, 730  
**Fibrosa obliterans**, iv, 595  
**Fibrosis in chronic myocarditis**, iv, 114  
   of lungs in bronchiectasis, ii, 917  
**Fibrous stenosis of œsophagus**, iii, 96  
**Filaria**, ii, 296-315  
   inflammation of thoracic duct and,  
     iv, 584  
**Filariasis**, ii, 305  
**Fingers in arthritis deformans**, v, 912  
   clubbing of, in bronchiectasis, ii,  
     916  
   in osteo-arthropathy, v, 984  
**Fish poisons**, ii, 451  
**Fissures of intima of aorta**, iv, 487  
**Fistula-in-ano in tuberculosis**, i, 432  
**Fistule, biliary, in cholelithiasis**, iii, 584  
**Fleas**, ii, 325  
**Fletcherism**, ii, 660  
**Flint murmur in aortic insufficiency**, iv,  
   226  
**Floating spleen**, iv, 938  
**Florida fever**, i, 1011  
**Fluke infections**, ii, 225  
**Fluoroscope in congenital disease of heart**,  
   iv, 444  
**Fluoroscopic findings in constipation**, iii,  
   313  
**Flutter, auricular**, iv, 86, 92, 94, 99  
**Folin's theory of protein metabolism**, ii,  
   594  
**Follicular pharyngitis**, ii, 845  
   ulcers, iii, 331  
**Foot, painful**, i, 760  
**Foot-jerk**, v, 77  
**Foot-and-mouth disease**, i, 1025  
**Foramen ovale, patent**, iv, 355  
**Foreign bodies in appendicitis**, iii, 360  
   in bronchi, ii, 926  
   intestinal obstruction from, iii,  
     404  
   protein, ii, 651  
**Fossa ovalis, aneurismal pouching of**, iv,  
   359  
**Fourth disease**, i, 915  
   ventricle, tumors of, v, 338  
**Fracture, intra-uterine**, v, 979  
**Fragilitas ossium**, v, 947, 980  
   congenital, v, 978  
**Fragmentation of myocardium**, iv, 116  
**Frenum of tongue, deformities of**, iii, 61  
   ulcer of, iii, 63  
**Freud's hypothesis of hysteria**, v, 657  
**Friedreich's ataxia**, v, 130  
**Fröhlich's syndrome**, iv, 815; v, 341  
**Frontal lobe, tumors of**, v, 331  
**Furuncles in scarlet fever**, i, 878  
   in typhoid fever, i, 101  
**Fusiform aneurism**, iv, 479

## G

- GALACTOSURIA, ii, 600
- Gall-bladder, anomalies of, iii, 547
- carcinoma of, iii, 593
- of liver and, iii, 543
- of pancreas and, iii, 652
- in catarrhal cholangitis, iii, 554
- in cholelithiasis, iii, 579
- in chronic peritonitis, iii, 719
- diseases of, peritonitis and, iii, 673
- empyema of, iii, 563, 566
- in suppurative cholangitis, iii, 557
- tuberculosis of, i, 364
- in typhoid fever, i, 86, 119
- wall of, biliary infection through, iii, 551
- Gall-ducts, tuberculosis of, i, 364
- Gall-stones, iii, 572
- chronic cholangitis and, iii, 559
- cholecystitis and, iii, 568
- colic, iii, 576
- infection and, iii, 578
- in common duct, iii, 581
- in cystic duct, iii, 580
- in diverticulum of Vater, iii, 582
- glycosuria and, ii, 678
- intestinal obstruction from, iii, 403
- Ganglia of cerebral hemisphere, v, 278
- in cerebrospinal meningitis, i, 598
- inflammation of, neuralgia and, v, 726
- Ganglionated neuroma of peritoneum, iii, 731
- Gangrene, acute arteritis and, iv, 450
- in aortic embolism, iv, 566
- thrombosis, iv, 566
- of appendix, iii, 363
- in cervical ribs, v, 556
- in chickenpox, i, 552
- in diabetes mellitus, ii, 705
- in diseases of cord, iv, 992
- in ergotism, iv, 995
- from x-rays, ii, 336
- in hemiplegia, iv, 992
- in infectious diseases, iv, 978, 993
- in lobar pneumonia, i, 217
- of lung, ii, 988
- in bronchiectasis, ii, 917
- in bronchopneumonia, ii, 964
- in fetid bronchitis, ii, 912
- in influenza, i, 541
- in lobar pneumonia, i, 251
- pneumothorax and, ii, 1074
- in tuberculosis, ii, 430
- tumors of lung and, ii, 1001
- in obliterative arteritis, iv, 993
- of pancreas, necrosis and, iii, 615
- in pathomimia, iv, 995
- post-vaccinal, i, 842
- in Raynaud's disease, iv, 983, 986
- in scarlet fever, i, 878
- of skin in smallpox, i, 812
- in thrombo-angiitis obliterans, iv, 547
- in typhoid fever, i, 101, 149
- Gangrenous appendicitis, iii, 372
- bronchitis, ii, 911
- cholecystitis, iii, 563
- Gangrenous pyelitis, iii, 954
- stomatitis, iii, 49
- Gas analysis in pneumothorax, ii, 1081
- Gaseous distension of intestines, iii, 298
- liver, iii, 466
- Gastralgia, nervous, iii, 135
- in mycotic gastritis, iii, 236
- Gastralgokenosis, iii, 134
- Gastric anacidity, iii, 153
- analysis in carcinoma of stomach, iii, 210
- in gastric ulcer, iii, 181
- atrophy, pernicious anemia and, iv, 623
- contents in atony of stomach, iii, 162
- in chronic gastritis, iii, 242
- crises in tabes dorsalis, v, 841, 846
- disease, carcinoma and, iii, 200
- tetany and, v, 749
- emptiness, painful, iii, 134
- hyperesthesia, iii, 137
- idiosyncrasies, iii, 138
- juice, hypersecretion of, iii, 122
- nerves, paralysis of, v, 523
- neurasthenia, iii, 164
- neuroses, iii, 106
- regurgitation, iii, 140
- tuberculosis, i, 388, 431
- ulcer, iii, 168
- chronic peritonitis and, iii, 714
- intestinal hemorrhage and, iii, 307
- pyloric stenosis and, iii, 189
- subphrenic abscess and, iii, 70
- pyopneumothorax and, iii, 710
- tetany and, iii, 189
- in tuberculosis, i, 432; iii, 170
- Gastritis, acute, iii, 226
- chronic, iii, 237, 240
- diphtheritic, iii, 236
- interstitial purulent, iii, 231
- mesenteric lymphadenitis and, iv, 736
- mucous, iii, 238
- mycotic, iii, 235
- phlegmonous, iii, 231
- proliferative, iii, 238
- secondary, iii, 239
- syphilitic, ii, 174
- toxic, iii, 233
- Gastrodiscus hominis, ii, 238
- Gastro-enteritis in lead poisoning, ii, 360
- Gastro-intestinal auto-intoxication, ii, 506, 511, 524
- cardiac neuroses, iv, 297
- complications in lobar pneumonia, i, 253
- disease, typhoid fever and, i, 72, 158
- influenza, i, 543
- sepsis, pernicious anemia and, iv, 623
- splanchnoptosis, iii, 754
- symptoms in amœbic dysentery, ii, 35
- tract in angioneurotic œdema, iv, 1007
- in cirrhosis of liver, iii, 517
- in diabetes mellitus, ii, 705
- in jaundice, iii, 455
- in measles, i, 906



- Gastro-intestinal tract in pernicious anemia, iv, 634
- Gastromyorrhea, iii, 130
- Gastropexia, iii, 754
- in atony of stomach, iii, 162
- gastric and, iii, 254
- Gastrosuccorhea, iii, 122
- continua chronica, iii, 125
- periodica, iii, 125
- digestive, iii, 128
- in pylorospasm, iii, 150
- Gaucher's splenomegaly, iv, 962
- Gelatinous pneumonia, i, 352
- Gelle's test, v, 24
- Geniculate ganglia, inflammation of, v, 505
- Genital organs in hypopituitarism, iv, 816
- Genito-urinary diagnosis, iii, 1026
- system in lead poisoning, ii, 365
- tract, acute endocarditis and, iv, 153
- in influenza, i, 545
- in plague, i, 629
- in tuberculosis, i, 365, 389
- in typhus fever, i, 926, 930
- Geographical tongue, iii, 63
- Gerlier's disease, v, 515
- Giant urticaria, iv, 998
- Gigantism, iv, 806
- achondroplasia and, v, 961
- Gigantorhynchus, ii, 297
- Glanders, i, 649
- Glands, lymphatic, in typhoid fever, i, 146
- mesenteric, in typhoid fever, i, 85
- in smallpox, i, 790
- submaxillary, in typhoid fever, i, 104
- Glandular enlargement in arthritis deformans, v, 909, 915
- in lymphoid leukemia, iv, 671
- in pernicious anemia, iv, 635
- fever, i, 1012
- swelling in Hodgkin's disease, iv, 765
- system in typhoid fever, i, 145
- tuberculosis, i, 412
- Glaucoma in congenital cyanosis, iv, 339
- in trigeminal neuralgia, v, 734
- Glénard's disease, iii, 747
- Gliadins in nutrition, ii, 646
- Glioma of brain, v, 284, 311, 312
- Gliomatosis in syringomyelia, v, 213
- Gliosis in epilepsy, v, 599
- Glossitis, iii, 62, 63
- in typhoid fever, i, 103
- Glossodynia, iii, 63, 64
- Glosso-labio-laryngeal paralysis, v, 113
- Glossopharyngeal nerve, diseases of, v, 516
- Glossoplegia, iii, 62
- Glucose in urine, iii, 821
- Gluteal nerves, paralysis of, v, 558
- Glyciphagus ii, 323
- Glycocele, ii, 741
- Glycosuria, ii, 543; iii, 821
- in acromegaly, iv, 812
- alimentary, ii, 690
- brain tumor and, ii, 678; v, 338, 342
- in calculus of pancreas, iii, 635
- in carcinoma of pancreas, iii, 652
- cerebral hemorrhage and, ii, 678
- diseases of pancreas and, iii, 604, 606
- Glycosuria in exophthalmic goitre, iv, 868
- gall-stones and, ii, 678
- in gangrenous pancreatitis, iii, 619
- in gout, ii, 750
- in interstitial pancreatitis, iii, 630
- in necrosis of pancreas, iii, 617
- in suppurative pancreatitis, iii, 625
- Glycuronic acid in metabolism, ii, 599
- in urine, iii, 823
- in diabetes mellitus, ii, 702
- Gnathostoma siamense, ii, 302
- Goitre, iv, 835
- aberrant, iv, 841
- acute, iv, 840
- cardiac insufficiency and, iv, 199
- chronic, iv, 840
- colloid, iv, 839
- exophthalmic, iv, 849
- abortive, iv, 870
- acute, iv, 870
- angioneurotic oedema and, iv, 1000
- cardiac insufficiency and, iv, 201
- chronic, iv, 870
- enlargement of thymus and, iv, 913
- hypertrophy of heart and, iv, 182
- incomplete, iv, 870
- iodine and, iv, 851, 857, 875
- thyroiditis and, iv, 829
- fibrous, iv, 840
- intralaryngeal, iv, 842
- intratracheal, iv, 842
- iodism and, iv, 823, 847
- myxedema and, iv, 890
- parenchymatous, iv, 839
- radium and, iv, 838
- vascular, iv, 839
- water and, iv, 838
- Gonococcal adenitis, i, 761
- arthritis, i, 753
- bone lesions, i, 760
- chondritis, i, 760
- conjunctivitis, i, 746
- neonatorum, i, 747
- cystitis, i, 762
- endocarditis, i, 750
- erythema, i, 764
- nodosum, i, 764
- hyperkeratosis, i, 765
- infections, i, 743
- meningomyelitis, i, 761
- myocarditis, i, 752
- myositis, i, 760
- neuralgia, i, 761
- neuritis, i, 761
- pericarditis, i, 752
- perichondritis, i, 760
- peritonitis, i, 763; iii, 686
- phlebitis, i, 752
- polyneuritis, i, 761
- proctitis, i, 747
- sciatica, i, 761
- septicemia, i, 749
- stomatitis, i, 748; iii, 51
- tenosynovitis, i, 760
- urticaria, i, 764

- Gonococcal wound infection, i, 748  
 Gonococcal infections, chronic peritonitis and, iii, 714  
     kidney infections and, iii, 952  
     tuberculosis of kidney and, iii, 978  
 Gonococcus, i, 743  
     chronic nephritis and, iii, 880  
 Gonorrhoea, endocarditis and, iv, 153, 157  
     lobar pneumonia following, i, 267  
     myocarditis and, iv, 108  
     pericarditis and, iv, 42  
     typhoid fever and, i, 158  
 Gonorrhoeal bubo, iv, 739  
     heel, i, 760  
 Gordius ii, 296  
 Gout, ii, 729  
     acute, ii, 745  
     alcohol and, ii, 730  
     arthritis deformans and, v, 930  
     asthma and, ii, 933  
     chronic, ii, 746  
     diabetes mellitus and, ii, 677, 750  
     lead poisoning and, ii, 731  
     migraine and, v, 720  
     phlebitis and, iv, 574  
     trauma and, ii, 732  
 Graefe's sign in exophthalmic goitre, iv, 863  
 Granular casts, iii, 799  
     pharyngitis, ii, 845  
     pyelitis, iii, 954  
 Granuloma of brain, infectious, v, 319  
     of oesophagus, iii, 85  
 Gravel, sediment, iii, 1017  
 Graves' disease, iv, 849  
 Grocers' itch, ii, 323  
 Groin glands, varicose, ii, 308  
 Guinea-worm infection, ii, 303  
 Gumma of arteries, ii, 181  
     of brain, v, 320  
     of heart, ii, 183  
     of kidney, ii, 180  
     of liver, ii, 176  
     of lung, ii, 172  
     of spleen, ii, 176  
     of stomach, ii, 174  
     in syphilis, ii, 150  
     of trachea, ii, 170  
 Gummatous syphiloderma, ii, 184  
 Gums in infantile scurvy, ii, 806  
     in scurvy, ii, 806  
     in tuberculosis, i, 387; iii, 57  
     in ulcerative stomatitis, iii, 48
- ## H
- HABIT chorea, v, 784  
     spasm, v, 784  
 Hæmonchus contortus, ii, 294  
 Hallucinations, v, 37, 39  
 Halzoun, ii, 236  
 Hamularia equina, ii, 315  
 Hands in achondroplasia, v, 962  
     in aneurism of aorta, iv, 495  
     in arthritis deformans, v, 912  
     in myxoedema, iv, 891  
     in osteo-arthropathy, v, 981  
 Hanot's cirrhosis, iii, 535  
 Harelip, iii, 45  
 Harris' immunization in rabies, i, 971  
 Harvest mites, ii, 321  
 Hay asthma, ii, 860  
     fever, ii, 855  
     American, ii, 859  
 Head, actinomycosis of, i, 1038  
     sweating in rickets, ii, 782  
     tetanus, i, 579  
 Headache, sick, v, 709  
 Heart, abdominal, iv, 346  
     in acute myocarditis, iv, 116  
     in Addison's disease, iv, 789, 793  
     air in, cardiac insufficiency and, iv, 194  
     in alcoholism, ii, 401, 403  
     in amyloid kidney, iii, 935  
     aneurism of, iv, 482  
         of aorta and, iv, 499  
         in myocarditis, iv, 115, 127  
     anomalies of, iv, 350  
     in aortic insufficiency, iv, 223  
     apex of, bifid, iv, 350  
     arrhythmia of, iv, 29  
     in arterial transposition, iv, 386  
     in arteriosclerosis, iv, 462  
     beat, alternation of, iv, 88  
         mechanism of, iv, 78  
         pathological, iv, 92  
     premature contractions in, iv, 84  
     rapid, iv, 81  
     respiratory irregularity and, iv, 83  
     slow, iv, 82  
     biloculate, iv, 370  
     cervical, iv, 346  
     chambers of, displacement of, iv, 350  
     in chlorosis, iv, 651  
     in chronic nephritis, iii, 884, 898  
     in cirrhosis of liver, iii, 518, 525  
     in coarctation of aorta, iv, 427, 431  
     in congestion of liver, iii, 481  
     development of, iv, 323  
     diastole of, iv, 22, 32  
     digestive disturbances and, iv, 310  
     dilatation of, iv, 30, 187, 189  
     in diphtheria, i, 694, 702  
     diseases of, congenital, iv, 323  
         convulsions and, v, 794  
         neuroses and, iv, 296  
     displacements of, iv, 346  
     diverticulum of, iv, 351  
     in exophthalmic goitre, iv, 860  
     fatty, in myocarditis, iv, 115  
     fibrillation of, insufficiency and, iv, 192  
     functional diseases of, iv, 275  
     gastric distension and, iv, 311  
     in goitre, iv, 843  
     in gout, ii, 749  
     gumma of, ii, 183  
     hypertrophy of, iv, 30, 166  
         congenital, iv, 351  
         defects of interauricular septum and, iv, 359

- Heart, impulse of, iv, 21  
 insufficiency of, iv, 187, 191  
   acute, iv, 191  
   chronic, iv, 198  
   hemopericardium and, iv, 193  
   right, iv, 197  
 irritable, iv, 319  
 in jaundice, iii, 453  
 in lobar pneumonia, i, 226  
 in lymphoid leukemia, iv, 671  
 mechanism of, iv, 27, 278, 314  
 muscle, degeneration of, iv, 190  
   functions of, iv, 78, 187  
   reserve power of, iv, 188, 216  
   tonicity of, iv, 190  
 in myxedema, iv, 894  
 pectoral, iv, 346  
 in pericarditis, iv, 45, 48  
 in pernicious anemia, iv, 632  
 in plague, i, 628  
 in Raynaud's disease, iv, 990  
 in rheumatic fever, i, 984  
 rupture of, hemopericardium and, iv, 70  
   insufficiency and, iv, 193  
   in myocarditis, iv, 115, 127  
 in scarlet fever, i, 865, 877  
 senile, myocarditis and, iv, 126  
 septum of, absence of, iv, 370  
   anomalies of, iv, 352  
   development of, iv, 370  
 in serofibrinous pleuritis, ii, 1022  
 sexual organs and, iv, 316  
 standstill of, iv, 83  
 in Stokes-Adams syndrome, iv, 307  
 strain, iv, 82  
   myocarditis and, iv, 109  
 syphilis of, ii, 183  
   of myocardium and, iv, 143  
 in toxemia, i, 563  
 triloculate, iv, 372  
 in tuberculosis, i, 385, 433  
 in typhoid fever, i, 125  
 valves of, aneurism of, iv, 482  
   lesions of, combined, iv, 259  
   rupture of, aortic insufficiency and, iv, 219, 227  
   insufficiency and, iv, 193  
 weakness, iv, 319  
 work of, iv, 33  
 wounds of, insufficiency and, iv, 191
- Heart-block, iv, 83  
 in lobar pneumonia, i, 257  
 Stokes-Adams syndrome and, iv, 306  
 ventricular systole and, iv, 89
- Hearts, multiple, iv, 350
- Heat, burns from, ii, 348  
 exhaustion, ii, 330
- Heberden's nodes, v, 921
- Heel, gonorrhoeal, i, 760
- Helminthoma elasticum, ii, 308
- Hemangioma of intestines, iii, 434
- Hemarthrosis in hemophilia, iv, 350
- Hematocnesis, iii, 279
- Hematochyluria bilharzian, ii, 239  
 in filariasis, ii, 309
- Hematogenous jaundice, iii, 447
- Hematology, main data of, iv, 605
- Hematoma in hemophilia, iv, 723  
 retroperitoneal, iii, 738
- Hematomyelia, v, 260  
 in cervical enlargement, v, 262  
 in conus medullaris, v, 262  
 in lumbosacral enlargement, v, 262  
 in purpura hemorrhagica, iv, 712
- Hematopoiesis in spleen, iv, 934
- Hematoporphyrinuria, iii, 809
- Hematorrachis, v, 258  
 in purpura hemorrhagica, iv, 712
- Hematuri, iii, 802  
 in carcinoma of prostate, iii, 1038  
 in chronic nephritis, iii, 901  
 Egyptian, ii, 239  
 in embolism, iv, 558  
 endemic, ii, 239  
 in Henoch's purpura, iv, 710  
 in hypernephroma, iii, 996  
 in infantile scurvy, ii, 806  
 in nephroptosis, iii, 756  
 in polymyositis hemorrhagica, v, 879  
 in purpura hemorrhagica, iv, 711  
 in renal infarction, iv, 564  
 in sarcoma of kidney, iii, 1003  
 in tuberculosis of kidney, iii, 984  
 in tumors of kidney, 1005  
 in typhoid fever, i, 143
- Hemeralopia, v, 35, 480
- Hemichromatopsia in brain tumors, v, 334
- Hemiambyopia, v, 34
- Hemianesthesia in hysteria, v, 681  
 in lesion of internal capsule, v, 276  
 of pons, v, 290
- Hemianopic pupillary reaction, v, 482
- Hemianopsia, v, 34, 270, 480  
 in brain abscess, v, 381, 382, 384  
 tumor, v, 332, 334  
 in hemiplegia, v, 468  
 heteronyma, v, 34  
 homonyma, v, 34  
 in lesion of internal capsule, v, 276  
 quadrant, v, 34
- Hemiataxia in abscess of brain, v, 381, 384
- Hemiathectosis, v, 70
- Hemiatrophy, facial, v, 991
- Hemicardia, v, 350
- Hemichromatopsia, v, 34
- Hemicontracture in lesion of internal capsule, v, 277
- Hemicrania, v, 709
- Hemihyperesthesia in hysteria, v, 681
- Hemiparesis in abscess of brain, v, 384
- Hemiparesthesia in migraine, v, 712
- Hemiplegia in acute miliary tuberculosis, i, 421  
 apoplexy and, v, 412, 457  
 cerebral, congenital, v, 409  
 in diabetes mellitus, ii, 707  
 hysteria and, v, 421, 678  
 infantile, v, 412  
 in lesion of internal capsule, v, 276  
   of optic thalamus, v, 279  
   of pons, v, 290  
   of pontomedullocerebellar space, v, 290

- Hemiplegia in lobar pneumonia, i, 262  
 Hemochromatosis, ii, 681; iii, 529, 606  
   interstitial pancreatitis and, iii, 630  
   portal cirrhosis of liver and, iii, 510  
 Hemofuscin in liver, iii, 470  
 Hemoglobin, estimation of, iv, 607  
 Hemoglobinuria, iii, 818  
   in angioneurotic oedema, iv, 1008  
   in blackwater fever, ii, 113  
   in Raynaud's disease, iv, 989  
 Hemohydrothorax, ii, 1054  
 Hemolymph nodes, iv, 613, 753  
   in Banti's disease, iv, 753  
   in leukemia, iv, 753  
   in malarial fever, iv, 754  
   in mercuric chloride poisoning, iv, 754  
   in pernicious anemia, iv, 753  
   in posthemorrhagic anemia, iv, 753  
   in pseudomelanosis, iv, 753  
   in trypanosomiasis, iv, 754  
 Hemolysis, secondary anemia and, iv, 655  
   in spleen, iv, 934  
 Hemopericardium, iv, 70  
   cardiac insufficiency and, iv, 193  
 Hemoperitoneum, iii, 669  
 Hemophilia, iv, 717  
 Hemopneumopericardium, iv, 71  
 Hemopoietic tissues, atypical, sarcoma-like growth of, iv, 679  
 Hemoptysis, ii, 951  
 Hemorrhage, cerebral, v, 401, 410, 411, 414, 415  
   extradural, v, 257  
   intestinal, iii, 307  
   intradural, v, 258  
   mediastinal, ii, 1110  
   into spinal cord, v, 260  
   membranes, v, 257  
 Hemorrhagic chickenpox, i, 852  
   cysts of spleen, iv, 951  
   diarrhœa in embolism, iv, 558  
   diathesis in vaccination, i, 842  
   erosions of stomach in uremia, iii, 838  
   exanthemata in syphilis, ii, 189  
   exudate in pericarditis, iv, 44  
   necrosis of pancreas, iii, 610  
   pericarditis, iv, 50  
   pleural fluids, ii, 1053  
   pleurisy, ii, 1053  
   pustular smallpox, i, 804  
   septicemia, i, 558  
   typhoid fever, i, 153  
 Hemorrhoids, intestinal hemorrhage and, iii, 307  
 Hemoserothorax, ii, 1053  
 Hemosiderin in liver, iii, 470  
   in malarial fever, ii, 68  
 Hemothorax, ii, 1054  
 Henoch's purpura, iv, 708  
 Hepatic artery, abscess of liver and, iii, 497  
   aneurism of, iii, 458  
   dilatation of, iii, 458  
   embolism of, iii, 458; iv, 569  
   thrombosis of, iii, 458  
   distomatosis, ii, 229  
 Hepatic infarction, iv, 569  
   intermittent fever, iii, 582  
   splachnoptosis, iii, 754  
   toxemia, myocarditis and, iv, 124  
   veins, abscess of liver and, iii, 497  
   dilatation of, iii, 459  
   embolism of, iii, 460  
   thrombosis of, iii, 460; iv, 545  
 Hepatitis, acute, iii, 494  
   chronic interstitial, iii, 505  
   suppurative, iii, 496  
 Hepatogenous jaundice, iii, 447  
 Hepatoptosis, iii, 458, 757  
 Hepatopulmonary abscess, ii, 43, 48  
 Hereditary cerebellar ataxia, Marie's, v, 130, 236  
   spastic spinal paralysis, v, 105  
 Hernia, foraminal, of medulla, v, 324  
   internal, intestinal obstruction from, iii, 405  
   intestinal hemorrhage and, iii, 307  
   mediastinal, ii, 1109  
   of pericardium, iv, 346  
 Herpes in arsenic poisoning, ii, 375  
   buccalis, iii, 51  
   in influenza, i, 545  
   labialis, iii, 45  
   lymphangitis and, iv, 592  
   of œsophagus, iii, 90  
   in pneumonia, i, 232, 424  
   in typhoid fever, i, 90, 101  
   zoster, v, 571  
   of auricle, v, 505  
   in chorea, v, 781  
   in migraine, v, 719  
   in tuberculosis, i, 393, 435  
   in tumors of lung, ii, 1000  
 Herpetetic stomatitis, iii, 51  
 Herpetiform scleroderma, iv, 1024  
 Heterophytes heterophytes, ii, 238  
 Heterotaxy, incomplete, iv, 349  
 Hexathyridium venanum, ii, 235  
 Hexosephosphate, metabolism of, ii, 562  
 Hip-joint in arthritis deformans, v, 913, 922  
 Hippocratic fingers in biliary cirrhosis of liver, iii, 539  
   in tuberculosis, i, 411  
 Hirudineæ, ii, 316  
 His, bundle of, development of, iv, 328  
 Histogaster spermaticus, ii, 323  
 Hodgkin's disease, iv, 751, 755  
 Hoffman's symptom, v, 71  
 Holothyrus coccinella, ii, 323  
 Homogentisic acid, ii, 655  
   in urine, iii, 824  
 Hookworm disease, ii, 271  
 Horner's complex, iii, 103  
 Hour-glass stomach, iii, 182  
 Human trypanosomiasis, ii, 119, 127  
 Hunger, sense of, v, 193  
 Hunterian chancre, ii, 154  
 Huntington's chorea, v, 787  
 Hutchinson's teeth, iii, 59  
 Hyaline casts, iii, 799  
   degeneration of lymph nodes, iv, 746  
   of myocardium, iv, 116

- Hyaline perihepatitis, iii, 488  
 Hyaloserousitis, iv, 60, 63, 64  
 Hydatid cyst of brain, v, 319  
   of liver, subpleuric pyopneumo-  
   thorax and, iii, 711  
   of pericardium, iv, 76  
   of peritoneum, iii, 733  
   of spleen, iv, 953  
   disease, ii, 264  
 Hydrocephalus, v, 351  
   acquired, v, 356  
   epilepsy and, v, 599  
   essential, v, 353  
   externus, v, 353  
   idiopathic, v, 353  
   internus, v, 353  
   in meningitis, i, 610, 618  
   in rickets, ii, 789  
   secondary, v, 356  
   in tumor of brain, v, 338  
 Hydromyelia, v, 210  
 Hydronephrosis, renal calculus and, iii,  
   1018  
   tuberculosis of kidney and, iii, 978  
 Hydropericardium, iv, 69  
 Hydrophobia, i, 956  
 Hydropericardium, iv, 71  
 Hydrothorax, ii, 1052  
   in aneurism of aorta, iv, 508  
   in mitral insufficiency, iv, 236  
 Hymenolepis diminuta, ii, 246, 254  
   lanceolata, ii, 246, 254  
   nana, ii, 245, 252  
 Hypacusis, v, 33  
 Hypalgia in tabes dorsalis, v, 838  
 Hyperacidity, iii, 112  
   in chronic gastritis, iii, 242  
   in duodenal ulcer, iii, 643  
   larval, iii, 121  
   ulcer of stomach and, iii, 114, 171  
 Hyperalgesia, v, 30, 33  
   in appendicitis, iii, 367  
   in diseases of peripheral nerves, v, 539  
   in herpes zoster, v, 572  
   hysteria and, v, 682  
 Hyperchlorhydria, iii, 112  
   in chlorosis, iv, 651  
 Hyperchondroplasia, v, 969  
 Hyperemia of adrenal glands, iv, 782  
   of brain, v, 392  
   hypertrophy of heart and, iv, 168  
   of kidney, iii, 788  
   in Raynaud's disease, iv, 983, 985  
   of spinal cord, v, 257  
 Hyperepinephry, iv, 779  
 Hyperesthesia, v, 28  
   in acute meningitis, v, 170  
   in beriberi, ii, 495  
   in cerebrospinal meningitis, i, 607  
   in embolism of arteries, iv, 568  
   in erythromelalgia, iv, 1028  
   gastrica, iii, 137  
   in hysteria, v, 681  
   of larynx, v, 522  
   in leprosy, i, 526  
   in occupation neuroses, v, 747  
   in pernicious anemia, iv, 631  
 Hyperesthesia in Raynaud's disease, iv,  
   987  
   in tabes dorsalis, v, 836  
   in thrombosis of arteries, iv, 568  
   in tuberculosis, i, 501  
 Hyperkeratosis, gonococcal, i, 765  
   of tongue, iii, 61  
 Hypermetropia, v, 36  
 Hypermnnesia, v, 53  
 Hypernephroma, iii, 994  
   of adrenal glands, iv, 784  
   tuberculosis and, iii, 978  
 Hyperopia, v, 36  
 Hyperostosis, v, 969  
   of skeleton with cyst-building, v, 978  
   of skull, v, 988, 989  
 Hyperplasia of adrenal glands, iv, 781  
   of thymus, iv, 912  
   tuberculosis of lymph nodes and, iv,  
   741  
 Hyperplastic bone-marrow, iv, 612  
   intestinal tuberculosis, iii, 390  
 Hyperpnea in cardiac insufficiency, iv, 206  
 Hyperptyalism, iii, 68  
 Hyperpyrexia in tetanus, i, 578  
 Hypersarcosis, iv, 596  
 Hypersecretion of gastric juice, iii, 122  
 Hyperthymia, v, 60  
 Hyperthyroidism, chlorosis and, iv, 648  
   myocarditis and, iv, 113  
 Hypertony of muscles, v, 68  
 Hypertrophic biliary cirrhosis, iii, 535  
   internal pachymeningitis, v, 166  
   osteopathies, v, 969  
   pulmonary osteo-arthropathy, i, 411  
   stenosis of pylorus, iii, 263  
 Hypertrophy of adrenal glands, iv, 781  
   of auricles, iv, 184, 185  
   cardiac, in myocarditis, iv, 121  
   of heart, iv, 30, 116  
     congenital, iv, 351  
     defects of interauricular septum  
       and, iv, 359  
   of muscles, v, 68  
   of pharyngeal tonsil, ii, 840  
   of prostate, iii, 1035  
     kidney infections and, iii, 952  
   of thymus, iv, 912  
   of ventricles, iv, 185  
 Hypesthesia in disease of spinal roots, v,  
   561  
   in tabes dorsalis, v, 838  
 Hypomycetic stomatitis, iii, 50  
 Hypochlorhydria, iii, 152  
 Hypochondriasis in constipation, iii, 314  
 Hypoepinephry, iv, 779  
 Hypogastric reflexes, v, 78  
 Hypoglossal nerve, diseases of, v, 530  
 Hypomnesia, v, 53  
 Hypophysis cerebri, iv, 803  
   in exophthalmic goitre, iv, 857  
   in myxedema, iv, 890  
 Hypopituitarism, iv, 815  
   in brain tumor, v, 342  
 Hypoplasia of adrenal glands, iv, 781  
   of aorta, iv, 431  
   aneurism and, iv, 478

- Hypoplasia of aorta, defects of interauricular septum and, iv, 359  
 Hypothyria, v, 61  
 Hypothyroidism, iv, 880  
 Hypotonia in lesion of cerebellum, v, 287  
   in tabes dorsalis, v, 839  
 Hypotony of muscles, v, 68  
 Hypotyalism, iii, 68  
 Hysteria, v, 654  
   adductor paralysis and, v, 521  
   angioneurotic œdema and, iv, 1001  
   astasia-abasia and, v, 680  
   dementia præcox and, v, 695  
   electrical shock and, ii, 341  
   exophthalmic goitre and, iv, 870  
   hemiplegia and, v, 419, 424, 678  
   humoralism and, v, 654  
   mythomania and, v, 659  
   pithiatism and, v, 659  
   psychotherapy and, v, 702  
   traumatic, v, 644, 691  
   in typhoid fever, i, 136  
 Hysterical angina pectoris, iv, 295  
   astasia-abasia, v, 815  
   disseminated sclerosis, v, 154  
   insanity, trauma and, v, 647
- I**
- ICHTHYOSIS buccalis, iii, 64  
 Icterus in blackwater fever, ii, 114  
   neonatorum, iii, 450  
   in purpura hemorrhagica, iv, 711  
 Ideas, associative, anomalies of, v, 51  
   autochthonous, v, 52  
   exaggerated, v, 52  
   flight of, v, 46  
   hyperdynamic, v, 52  
   imperative, v, 52  
   intensity of, anomalies of, v, 52  
 Ideation, anomalies of, v, 46  
 Idiocy, alcoholism and, ii, 407  
   amaurotic family, v, 863  
   in endemic cretinism, iv, 882  
   in sporadic cretinism, iv, 885  
 Idiopathic abscess of brain, v, 385  
   dilatation of colon, iii, 320  
   phlebitis, iv, 574  
   purpura, iv, 691  
   recurrent thrombophlebitis, iv, 531  
 Ileocolitis in measles, i, 906  
 Ileum, carcinoma of, iii, 428  
 Ileus, iii, 394  
 Illuminating gas poisoning, ii, 388  
 Illusions, v, 37, 38, 55  
 Imbecility, epilepsy and, v, 594  
 Immunity, i, 35, 46, 65  
 Impetigo in congenital syphilis, ii, 190  
   contagiosa in scarlet fever, i, 878  
   in vaccination, i, 843  
   varicellosa, i, 852  
 Inanition toxemia, myocarditis and, iv, 124  
 Incarceration of intestines, iii, 394, 406  
 Incoherence, v, 43, 51  
 Incompetence, myocardial, of obese, iv, 128  
 Incontinence of pylorus, iii, 159  
 Incoördination in syringomyelia, v, 221  
 Indian liver-fluke disease, ii, 234  
 Indicanuria, ii, 509, 667; iii, 811  
 Infantilism, v, 967  
 Infarction, embolism and, iv, 556  
   gastric ulcer and, iii, 172  
   hepatic, iv, 569  
   intestinal hemorrhage and, iii, 307  
   of kidney, iii, 789; iv, 564  
   of lung, ii, 954; iv, 563  
   pulmonary, iv, 563  
   of spleen, iv, 946  
   in typhoid fever, i, 122, 129  
 Infection, aerial, i, 21  
   bronchial, i, 27  
   droplet, i, 24  
   duct, i, 21  
   gastro-intestinal, i, 27  
   germinal, i, 28  
   mixed, i, 31  
   nasopharyngeal, i, 26  
   oral, i, 26  
   secondary, i, 31  
   water-borne, i, 24  
 Infectious agents, i, 18  
   arthritis, v, 896  
   jaundice, i, 1014  
 Infective myelitis, v, 229  
   œsophagitis, iii, 81  
 Inferior vena cava, thrombosis of, iv, 541  
 Infiltration, fatty, ii, 604  
 Inflammation of adrenal glands, iv, 783  
   of brain, v, 359  
   of geniculate ganglia, v, 505  
   of labial glands, iii, 45  
   of lymphatic glands, iv, 730  
   of mouth, iii, 45  
   of œsophagus, iii, 79  
   of optic nerve, v, 476  
   of parotid gland, iii, 69  
   of spleen, iv, 940  
   in syphilis, ii, 151  
   of thoracic duct, iv, 582  
   of thymus, iv, 928  
   of thyroid gland, iv, 825  
   of ureter, iii, 974  
 Influenza, i, 534  
   acute endocarditis and, iv, 158  
   arteriosclerosis and, iv, 454  
   arthritis deformans and, v, 901  
   cardiac arrhythmia and, iv, 288  
   chronic, i, 541  
   gastro-intestinal, i, 543  
   genito-urinary, i, 545  
   lobar pneumonia and, i, 207, 266, 542  
   lymphadenitis and, iv, 737  
   myocarditis and, iv, 108, 117  
   nephritis and, iii, 851  
   nervous, i, 542  
   neurasthenia and, v, 629  
   respiratory, i, 539  
   sporadic, i, 534  
   tuberculosis and, i, 321  
   typhoid fever and, i, 158  
   typical, i, 538  
 Influenzal encephalitis, v, 362  
 Inguinal glands, lymphadenitis of, iv, 736

- Inguinal glands, tuberculosis of, iv, 744  
 Inhalation pneumonia, i, 206  
 Innervation of lungs, ii, 814  
   of respiratory organs, ii, 814  
 Insanity, aneurism and, iv, 478  
   epileptic, trauma and, v, 647  
   hysterical, trauma and, v, 647  
   lobar pneumonia and, i, 208  
   in pellagra, ii, 485  
 Insects, parasitic, ii, 324  
 Insolation, ii, 331  
 Insufficiency, aortic, iv, 217  
   of heart, iv, 191  
   mitral, iv, 232  
     congenital, iv, 407  
   pulmonary, iv, 255  
   tricuspid, iv, 244  
     congenital, 407  
 Insular sclerosis, v, 140  
 Interacinar pancreatitis, iii, 629  
 Interauricular septum, defects of, iv, 355  
   development of, iv, 325  
 Intercostal neuralgia, v, 739  
 Interlobar empyema, ii, 1050  
 Interlobular pancreatitis, iii, 629  
 Intermittent claudication in arteriosclerosis, iv, 465  
   in thrombo-angitis obliterans, iv, 547  
   disseminated sclerosis, v, 151  
   gastromyorrhœa, iii, 130  
   œdema, iv, 998  
 Interstitial emphysema of lungs, ii, 982  
   hypertrophic progressive neuritis of childhood, v, 112  
   myocarditis, iv, 113, 114  
   pancreatitis, iii, 626  
 Interventricular septum, defects of, iv, 363  
   development of, iv, 327  
   undefeined space of, aneurism of, iv, 369  
 Intestinal anthrax, i, 645  
   colic, iii, 303  
   crises in tabes dorsalis, v, 842  
   disease, tetany and, v, 749  
   distomatosis, ii, 237  
   flora, relationships of, ii, 652  
   hemorrhage, iii, 307  
     in amœbic dysentery, ii, 44  
     in lobar pneumonia, i, 254  
     in tuberculosis, i, 419  
     in typhoid fever, i, 108  
   indigestion, iii, 301, 324  
   infection, arthritis deformans and, v, 903  
   parasites, anemia and, iv, 623, 641, 659  
     in lobar pneumonia, i, 254  
   plague, i, 635  
   processes, automatic mechanism in, iii, 291  
   roundworms, ii, 271  
   sand, iii, 295, 440  
   teniasis, ii, 245  
 Intestines, adenoma of, iii, 433  
   in amœbic dysentery, ii, 29  
   amyloid disease of, iii, 392  
   antiperistalsis of, iii, 287  
   arteriosclerosis of, iii, 437  
     in Asiatic cholera, i, 681  
     in bacillary dysentery, i, 772, 775  
   bacillus *aërogenes* in, iii, 296  
     *bifidus communis* in, iii, 296  
     *coli* in, iii, 296  
     *lactis aërogenes* in, iii, 296  
     *putrificus* in, iii, 296  
   bacteria in, iii, 295  
   carcinoma of, iii, 424  
     peritonitis and, iii, 426  
   compression of, iii, 394  
   constriction of, iii, 394  
   descent of, iii, 757  
   diseases of, peritonitis and, iii, 672  
   diverticula of, iii, 420  
   gaseous distension of, iii, 298  
   hemangioma of, iii, 434  
   human, amœba of, ii, 18  
   incarceration of, iii, 394  
   lipoma of, iii, 433  
   lymphangioma of, iii, 434  
   lymphosarcoma of, iii, 431  
   motor function of, iii, 286  
   movements of, iii, 287  
   myxoma of, iii, 433  
   neuralgia of, iii, 305  
   obstruction of, iii, 394  
     acute, iii, 396  
     in cholelithiasis, iii, 584  
     chronic, iii, 395, 397  
   obturation of, iii, 394  
   occlusion of, iii, 394  
   paralysis of, in peritonitis, iii, 681  
   perforation of, in gastric ulcer, iii, 186  
     in typhoid fever, i, 111  
   peristalsis of, iii, 286  
   rhythmic segmentation of, iii, 286  
   sarcoma of, iii, 431  
   in scarlet fever, i, 865  
   strangulation of, iii, 394  
   stricture of, iii, 394  
   syphilis of, ii, 175; iii, 391  
   tuberculosis of, i, 361, 432; iii, 387  
   tumors of, iii, 424, 433  
   in typhoid fever, i, 82, 105  
   ulcer of, iii, 343, 838  
 Intoxications, adrenal glands and, iv, 781  
   aneurism and, iv, 476  
   arteriosclerosis and, iv, 455  
   nephritis and, iii, 880  
   spleen and, iv, 936  
 Intracellular enzymes, ii, 663  
 Intracerebellar tumors, v, 338  
 Intracranial pressure, v, 391  
 Intradural hemorrhage, v, 258  
 Intralaryngeal goitre, iv, 842  
 Intra-ocular muscles, paralysis of, v, 486  
 Intrapertitoneal subphrenic abscess, iii, 703  
 Intrapleural pressure in pneumothorax, ii, 1072  
   tension in serofibrinous pleuritis, ii, 1014  
 Intratracheal goitre, iv, 842  
 Intra-uterine fracture, v, 979  
   rickets, v, 955  
 Intubation in diphtheria, i, 723

- Intussusception, intestinal obstruction  
     from, iii, 409  
     typhoid fever and, i, 159  
 Inverting enzymes, iii, 290  
 Iodide, purpura and, iv, 689  
 Iodine, exophthalmic goitre and, iv, 851, 857  
     thyroid gland and, iv, 823  
 Iodococcus vaginatus in mouth, iii, 40  
 Iodism, ii, 209  
 Irritability, pathological, v, 61  
 Irritable heart, iv, 319  
 Irritation, motor, v, 70  
 Ischemia of kidney, iii, 787  
     of myocardium, iv, 290  
     in Raynaud's disease, iv, 983  
 Ischiorectal abscess in tuberculosis, i, 432  
 Ischuria, iii, 791, 806  
 Islands of Langerhans, diabetes mellitus and, ii, 680, 693  
 Itch, grocer's, ii, 323  
     sarcoptic, ii, 318  
 Itching in uremia, iii, 840  
 Ixodiasis, ii, 317

## J

- JACKSONIAN epilepsy, v, 592  
 Jackstone calculus, iii, 1009  
 Japanese liver-fluke disease, ii, 230  
 Jaundice, iii, 446, 552  
     in abscess of liver, iii, 500  
     in acute cholangitis, iii, 553  
     cholecystitis, iii, 565  
     enteritis, iii, 330  
     gastritis, iii, 229  
     hepatitis, iii, 495  
     yellow atrophy of liver, iii, 475  
     in aneurism of hepatic artery, iii, 459  
     in biliary cirrhosis of liver, iii, 538  
     in calculus of pancreas, iii, 635  
     in carcinoma of gall-bladder, iii, 596  
     of liver, iii, 542  
     of pancreas, iii, 650, 652  
     catarrhal, iii, 448, 552  
     in cholelithiasis, iii, 579  
     in chronic cholangitis, iii, 560  
     peritonitis, iii, 714  
     chronic splenomegalic hemolytic, iii, 450  
     in cirrhosis of liver, iii, 520  
     of stomach, iii, 278  
     in congestion of liver, iii, 479, 484  
     in cyst of pancreas, iii, 645  
     in gangrenous pancreatitis, iii, 619  
     in gastritis, iii, 234  
     in Hodgkin's disease, iv, 766  
     hematogenous, iii, 447  
     hepatogenous, iii, 447  
     in hypernephroma, iii, 997  
     infectious, i, 1014  
     in lobar pneumonia, i, 259  
     in mitral insufficiency, iv, 236  
     nephritis and, iii, 854  
     in newborn, iii, 450  
     obstructive, iii, 447  
     in pernicious anemia, iv, 628

- Jaundice in portal thrombosis, iv, 544  
     in scarlet fever, i, 879  
     simple, iii, 448  
     in splenic anemia, iv, 958  
     in suppurative cholangitis, iii, 557  
     pancreatitis, iii, 624  
     in syphilis, ii, 158  
     in tuberculosis of peritoneum, iii, 726  
     in typhoid fever, i, 118  
     in typhus fever, i, 930  
 Jaw, actinomycosis of, iii, 59  
 Jejunal peptic ulcer, iii, 349  
 Jejunum, carcinoma of, iii, 428  
 Jigger flea, ii, 324  
 Joints in hemophilia, iv, 724  
     in purpura rheumatica, iv, 703  
     in Raynaud's disease, iv, 990  
     in scleroderma, iv, 1021  
     in syringomyelia, v, 225  
     tuberculosis of, i, 371  
 Jumping fleas, ii, 325  
 Juvenile vomiting, iii, 145

## K

- KAKKE, ii, 489  
 Kala-azar, iv, 964  
     acute hepatitis and, iii, 494  
 Keratitis in lobar pneumonia, i, 262  
 Keratosis in arsenic poisoning, ii, 375  
     buccalis, iii, 64  
 Kernig's sign in cerebrospinal meningitis  
     i, 599, 605; v, 170  
     in tuberculosis, i, 422  
 Ketone complex, ii, 546  
 Kidney, abscess of, iii, 937  
     in bronchiectasis, ii, 917  
     absence of, iii, 785, 937  
     acute dilatation of stomach and, iii, 251  
     in Addison's disease, iv, 795  
     adenoma of, iii, 1003  
     malignant papillary cyst, iii, 1002  
     in alcoholism, ii, 404  
     amyloid, iii, 933  
     tuberculosis and, iii, 933  
     anemia of, iii, 787  
     arteries of, anomalies of, iii, 786  
     arteriosclerotic, iii, 847, 897; iv, 463  
     in Asiatic cholera, i, 681  
     in blackwater fever, ii, 112  
     carcinoma of, iii, 1001  
     casts and, iii, 774  
     circulatory disturbances of, iii, 787  
     in cirrhosis of liver, iii, 518, 526  
     cystic congenital, iii, 786  
     in diabetes insipidus, ii, 722  
     mellitus, ii, 696  
     in diphtheria, i, 695  
     displacements of, iii, 786  
     excretion of solids by, iii, 768  
     of water by, iii, 767  
     in exophthalmic goitre, iv, 868  
     functional activity of, iii, 942  
     competence of, iii, 781  
     test of, iii, 781



- Kidney in gout, ii, 744  
 gumma of, ii, 180  
 in Hodgkin's disease, iv, 764  
 hyperemia of, iii, 788  
 hypernephroma of, iii, 994  
 infarct of, iii, 789  
 infections of, iii, 951; iv, 564  
 in influenza, i, 545  
 internal secretion of, iii, 769, 833  
 interstitial tissue of, in nephritis, iii, 859, 897  
 ischemia of, iii, 787  
 in lead poisoning, ii, 362  
 in malarial fevers, ii, 71  
 malformations of, iii, 785  
 mobility of, iii, 786  
 in nephritis, iii, 857, 881, 895  
 parathyroid glands and, iv, 904  
 pelvis of, tumors of, iii, 1006  
 phthalein test of, iii, 781  
 position of, anomalies of, iii, 786  
 of pregnancy, iii, 867  
 in renal calculus, iii, 1017  
 sarcoma of, iii, 1003  
 in scarlet fever, i, 866  
 secondary contracted, iii, 882, 884  
 shape of, anomalies of, iii, 786  
 in smallpox, i, 792  
 surgical, iii, 937  
 syphilis of, ii, 179  
 toxins and, iii, 773  
 tuberculosis of, i, 365; iii, 976, 979, 981  
   pyogenic infections and, iii, 952  
 tubules of, in acute nephritis, iii, 859  
   in chronic nephritis, iii, 856  
   disturbances of, iii, 772  
 tumors of, iii, 994, 1003, 1005  
 in typhoid fever, i, 86  
 variegated, iii, 882
- Kinanesthesias, v, 30  
 Klumpke palsy, v, 552  
 Knee-jerk, v, 77  
   in tabes dorsalis, v, 837  
 Knee-joint in arthritis deformans, v, 913, 923  
 Koch's bacillus emulsion, i, 305  
   tuberculin R., i, 305  
 Koplik spots in measles, i, 900  
 Kopp's asthma, iv, 918  
 Korsakow's psychosis, ii, 427; v, 45  
   alcoholism and, ii, 406, 427  
 Kussmaul's sign in adherent pericardium, iv, 62
- L**
- LABIAL glands, inflammation of, iii, 45  
 Labyrinth, choked, in brain tumor, v, 328  
 Lacrimal glands, enlargement of, iii, 73  
 Lactescent ascites, iii, 662  
 Lactosuria, ii, 600; iii, 822  
 Lancet fluke infection, ii, 233, 234  
 Landry's paralysis, v, 247  
 Language, zone of, v, 293  
 Larval hyperacidity, iii, 121  
 Laryngeal diphtheria, i, 698  
   muscles, spasm of, v, 522  
 Laryngeal nerve, paralysis of, v, 518  
   in patent ductus arteriosus, iv, 418  
   in thoracic aneurism, iv, 502  
   tuberculosis, i, 412  
 Laryngismus stridulus, ii, 880  
 Laryngitis, acute, ii, 868  
   chronic, ii, 870  
   in influenza, i, 537, 540  
   in lobar pneumonia, i, 223, 248  
   in measles, i, 906  
   oedematous, ii, 878  
   in rheumatic fever, i, 996  
   syphilitic, ii, 871  
   tuberculous, i, 348; ii, 876  
 Larynx in acromegaly, iv, 810  
   in angioneurotic oedema, iv, 1007  
   in disease of vagus nerve, v, 517  
   in hemiplegia, v, 459  
   in myasthenia gravis, v, 804  
   obstruction of, ii, 816  
   oedema of, ii, 878  
     in angioneurotic oedema, iv, 1005, 1007  
     lobar pneumonia and, i, 248  
   paralysis of, ii, 867  
   sensory neuroses of, v, 522  
   syphilis of, ii, 871  
   tuberculosis of, i, 348; ii, 876  
   in typhoid fever, i, 121  
 Lasègue's sign in sciatica, v, 563  
 Lateral sclerosis, v, 101  
 Lathyrism, v, 139  
 Lead colic, iii, 306  
   poisoning, ii, 355  
     amyotrophic lateral sclerosis and, v, 89  
     arteriosclerosis and, iv, 455  
     in children, ii, 369  
     secondary anemia and, iv, 658  
 Lecithins, metabolism of, ii, 562  
 Legs, hereditary oedema of, iv, 1011  
 Leishmania, iv, 964  
 Leishmaniasis, iv, 964  
 Lenticular degeneration, v, 278  
   nucleus, lesion of, v, 278  
   zone, lesion of, v, 278  
 Leontiasis ossea, v, 988  
 Leprosy, i, 522  
   of bronchi, ii, 906  
   of lymph nodes, iv, 746  
   of mouth, iii, 59  
   nerve, i, 527  
   tubercle, i, 528  
 Leptodera pellio, ii, 316  
 Leptomeninges in syphilitic myelitis, v, 239  
 Leptomeningitis, v, 167  
 Leptothrix buccalis in mouth, iii, 40  
 Leptus americanus, ii, 321  
   autumnalis, ii, 321  
   irritans, ii, 321  
 Leukanemia, iv, 682  
 Leukemia, iv, 659  
   acute, iv, 674, 677  
   atypical, iv, 679, 682  
   lymphoid, iv, 670

- Leukemia, mixed, iv, 679  
     myeloid, iv, 661  
 Leukemic blood without leukemic change  
     in blood-making organs, iv, 680  
     infiltration of liver, iii, 471  
     lymphocytoma, iv, 748  
     ulcers, iii, 352  
 Leukoblastic bone-marrow, iv, 611  
 Leukocytes in chlorosis, iv, 652  
     in Hodgkin's disease, iv, 769  
     in lymphoid leukemia, iv, 673  
     in myeloid leukemia, iv, 667  
     in pernicious anemia, iv, 638  
     in polycythemia, iv, 685  
     in splenic anemia, iv, 957  
     in tuberculosis, i, 383  
     in typhoid fever, i, 130  
 Leukocytosis, i, 40; iv, 614  
     in abscess of liver, iii, 502  
     in acute cholecystitis, iii, 565  
         meningitis, v, 171  
     in appendicitis, iii, 369  
     in arthritis deformans, v, 916  
     in cerebrospinal meningitis, i, 605  
     in erysipelas, i, 588  
     in fat embolism, iv, 560  
     in gangrenous pancreatitis, iii, 618  
     in Henoch's purpura, iv, 710  
     in infections, iv, 614  
     in inflammation of thoracic duct, iv, 583  
     in lobar pneumonia, i, 234  
     in lymphadenitis simplex, iv, 731  
     in necrosis of pancreas, iii, 617  
     in purpura, iv, 700  
     in rubella, i, 914  
     in smallpox, i, 808  
     in subphrenic pyopneumothorax, iii, 712  
     in suppurative cholangitis, iii, 558  
         pancreatitis, iii, 625  
     in tetanus, i, 578  
     toxic, iv, 614  
     in tuberculosis, i, 419  
         of peritoneum, iii, 726  
     in venous thrombosis, iv, 539  
     in whooping cough, i, 659  
 Leukoderma, ii, 186  
     in scleroderma, iv, 1019  
 Leukolysis in lobar pneumonia, i, 235  
 Leukoplakia, iii, 64  
 Leucosuria, ii, 600; iii, 821  
     in hepatic disease, iii, 445  
 Lichen planus of mouth, iii, 54  
     scrofulosum, i, 375  
 Lid reflex, v, 78  
 Liebig's theory of protein metabolism, ii, 591  
 Ligaments in rickets, ii, 782  
 Limnatis nilotica, ii, 316  
 Lineæ atrophicae in typhoid fever, i, 102  
 Lingua nigra, iii, 61  
 Linguatula serrata, ii, 323  
 Linguopapillitis, iii, 62  
 Linitis, plastic, iii, 273  
 Lipase, iii, 289  
 Lipemia in diabetes mellitus, ii, 695  
 Lipins, ii, 605  
 Lipogenous diabetes, ii, 676  
 Lipoids, ii, 605, 648  
 Lipoma of brain, v, 317  
     of intestines, iii, 433  
     of lung, ii, 998  
     of peritoneum, iii, 732  
     of thymus, iv, 932  
 Lipomatosis multiple, v, 818  
     universalis, ii, 761  
 Lips, actinomycosis of, iii, 59  
     double, iii, 44  
     eczema of, iii, 45  
     eruptions of, iii, 45  
     fissure of, iii, 44  
     tuberculosis of, iii, 57  
     tumors of, iii, 45  
 Lithemia, ii, 747  
 Lithemic splanchnoptosis, iii, 754  
 Lithiasis, bronchopulmonary, ii, 925  
 Lithosis, ii, 976  
 Lithuria, iii, 712  
 Litten's sign in pleural effusion, ii, 1023  
 Little's disease, v, 419  
 Liver, abscess of, iii, 496  
     in amoebic dysentery, ii, 38  
     subphrenic abscess and, iii, 700  
     in typhoid fever, i, 118  
     actinomycosis of, iii, 541  
     in acute hepatitis, iii, 495  
     acute yellow atrophy of, iii, 472  
     in alcoholism, ii, 403  
     amyloid, iii, 469  
         ascites and, iii, 658  
     angioma of, iii, 546  
     anomalies of, iii; 455, 456, 457  
     atrophy of, iii, 465  
     in blackwater fever, ii, 113  
     in bronchiectasis, ii, 917  
     carcinoma of, iii, 541  
         ascites and, iii, 542, 658  
     cardiac, iii, 481  
     in catarrhal cholangitis, iii, 554  
     in chronic interstitial nephritis, iii, 899  
         perihepatitis, iii, 490, 492  
         peritonitis, iii, 718  
     cirrhosis of, iii, 505  
         arthracotic, iii, 531  
         ascites and, iii, 658  
         biliary, iii, 535  
             alcohol and, iii, 536  
         Budd's non-alcoholic, iii, 509  
         in children, iii, 531  
         experimental, iii, 510  
         hemochromatosis and, iii, 529  
         intestinal hemorrhage and, iii, 307  
         in lenticular degeneration, v, 278  
         malarial, iii, 530  
         mesenteric thrombosis and, iii, 435  
         osteo-arthropathy and, v, 982  
         portal, iii, 508  
         tuberculosis of peritoneum and, iii, 526, 721  
     in congenital syphilis, ii, 190  
     congestion of, iii, 478, 481  
     constricted, iii, 456

- Liver**, cysts of, iii, 547  
     in defects of interauricular septum, iv, 362  
     deformities of, iii, 456  
     diabetes mellitus and, ii, 681, 685, 696  
     disorders of, retrogressive, iii, 465  
     emphysematous, iii, 466  
     enlargement of, in lobar pneumonia, i, 260  
     fatty, iii, 466  
     function of, iii, 31, 444  
         disorders of, iii, 442  
     gaseous, iii, 466  
     in Gaucher's splenomegaly, iv, 963  
     gumma of, ii, 176  
     in hepatic thrombosis, iv, 546  
     in Hodgkin's disease, iv, 763, 767  
     in jaundice, iii, 454  
     in kala-azar, iv, 970, 972  
     leukemic infiltration of, iii, 471  
     in lymphoid leukemia, iv, 672  
     in malarial fevers, ii, 70, 73  
     in mitral insufficiency, iv, 236, 237  
     in myeloid leukemia, iv, 663, 666  
     necrosis of, iii, 471  
     parenchymatous degeneration of, iii, 466  
     pericarditic pseudocirrhosis of, iii, 488, 716; iv, 60  
     in pernicious anemia, iv, 627, 634  
     pigmentation of, iii, 470  
     in plague, i, 629  
     in polycythemia, iv, 685  
     sarcoma of, iii, 546  
     in splenic anemia, iv, 955  
     sugar-iced, iii, 488, 718  
     in suppurative cholangitis, iii, 557  
     syphilis of, ii, 176; iii, 541  
         chronic peritonitis and, iii, 717  
     in thrombosis of portal vein, iii, 462  
     in tricuspid stenosis, iv, 253  
     tropical, iii, 494  
     tuberculosis of, i, 363; iii, 541  
     tumors of, iii, 541, 546  
     in typhoid fever, i, 85, 118
- Liver-fluke disease**, ii, 229
- Lobar pneumonia**, i, 202
- Lobstein's disease**, v, 980
- Lobulated spleen**, iv, 938
- Lockjaw**, i, 575
- Locomotor ataxia**, v, 833
- Loculated tuberculosis of peritoneum**, iii, 723
- Logorrhœa in aphasia**, v, 297
- Lordosis in acromegaly**, iv, 810
- Lousiness**, ii, 326
- Ludwig's angina**, iii, 75  
     cervical lymphadenitis and, iv, 733  
     in scarlet fever, i, 878
- Lucs venera**, ii, 144
- Luetin**, ii, 193
- Lumbago**, v, 644, 945
- Lumbar plexus, neuralgia of**, v, 739  
     paralysis of, v, 560  
     puncture, i, 608, 612; v, 171, 174
- Lumbo-abdominal neuralgia**, v, 739
- Lump jaw**, i, 1033
- Lung**, abscess of, ii, 993  
     in bronchopneumonia, ii, 964  
     in influenza, i, 538, 540  
     in lobar pneumonia, i, 250  
     pneumothorax and, ii, 1074  
     in tuberculosis, i, 430
- adenoma of, ii, 997
- in alcoholism, ii, 403
- in aneurism of aorta, iv, 503, 507
- arteries of, obliteration of, cardiac insufficiency and, iv, 195
- brown induration of, ii, 945
- carcinoma of, ii, 998  
         of liver and, iii, 543
- circulatory disturbances of, ii, 945
- cirrhosis of, ii, 969  
         of liver and, iii, 518
- congestion of, ii, 945
- in defects of interauricular septum, iv, 362
- diphtheria of, i, 700
- embolism of, ii, 954; iv, 560, 563
- emphysema of, ii, 979
- enchondroma of, ii, 997
- fibroma of, ii, 998
- fibrosis of, in bronchiectasis, ii, 917
- gangrene of, ii, 988  
         in bronchiectasis, ii, 917  
         in bronchopneumonia, ii, 964  
         in fetid bronchitis, ii, 912  
         foreign bodies and, ii, 989  
         in influenza, 541  
         in lobar pneumonia, i, 251  
         pneumothorax and, ii, 1074  
         in scurvy, ii, 802  
         in tuberculosis, i, 430
- gumma of, ii, 172
- in Hodgkin's disease, iv, 764
- infarction of, ii, 954; iv, 563
- innervation of, ii, 814
- lipoma of, ii, 998
- mechanism of, ii, 826
- new growths of, ii, 997
- œdema of, ii, 948  
         in serofibrinous pleuritis, ii, 1026
- osteoma of, ii, 997
- perforation of, in pleuritis, ii, 1026, 1044
- sarcoma of, ii, 998
- in smallpox, i, 790
- syphilis of, ii, 171
- tricuspid insufficiency and, iv, 246
- tuberculosis of, i, 348  
         bronchial glands and, iv, 743  
         pericarditis and, iv, 42
- tumors of, ii, 997
- in typhoid fever, i, 80
- ulceration of, in fetid bronchitis, ii, 912
- Lung-fluke disease**, ii, 226
- Lupus**, i, 374  
     erythematous of mouth, iii, 53
- Lymph cysts**, iv, 586  
     of spleen, iv, 951
- fistula, iv, 595, 598
- glands in anæmic dysentery, ii, 32  
         in diphtheria, i, 695  
         inflammation of, iv, 730

- Lymph glands in lymphoid leukemia, iv, 671  
 in myeloid leukemia, iv, 663, 666  
 in plague, i, 624  
 relation of, to blood, iv, 613  
 in rubella, i, 915  
 in scarlet fever, i, 866, 877  
 in sleeping sickness, ii, 129  
 tuberculosis of, i, 257  
 in typhoid fever, i, 146  
 nodes, amyloid degeneration of, iv, 746  
 calcification of, iv, 746  
 carcinoma of, iv, 747  
 cysts of, iv, 753  
 degeneration of, iv, 746  
 endothelioma of, iv, 747  
 in enlargement of thymus, iv, 916  
 in exophthalmic goitre, iv, 867  
 in Gaucher's splenomegaly, iv, 963  
 hematogenous affections of, iv, 730  
 in Hodgkin's disease, iv, 756  
 infiltration of, iv, 746  
 inflammation of, iv, 730  
 leprosy of, iv, 746  
 in leukemia, iv, 663, 666, 671, 747, 751  
 neoplasms of, iv, 747, 752  
 parasites of, iv, 753  
 in pernicious anemia, iv, 627  
 pigmentation of, iv, 746  
 sarcoma of, iv, 747  
 scrofula of, iv, 745  
 in splenic anemia, iv, 956  
 syphilis of, iv, 745  
 tuberculosis of, iv, 741  
 tumors of, iv, 747, 752  
 scrotum in filariasis, ii, 309  
 tissue, tuberculosis of, i, 357  
 varices in filariasis, ii, 309  
 vessels, carcinoma of, iv, 601  
 chondrosarcoma of, iv, 601  
 endothelioma of, iv, 600, 601  
 lymphangioma of, iv, 600  
 lymphosarcoma of, iv, 601  
 neoplasms of, iv, 600, 601  
 sarcoma of, iv, 601  
 syphilis of, iv, 596  
 tuberculosis of, iv, 595
- Lymphadenitis, iv, 730  
 in amoebic dysentery, iv, 738  
 in anthrax, iv, 737  
 of axillary glands, iv, 735  
 of bronchial glands, iv, 735  
 in bubonic plague, iv, 740  
 in cerebrospinal meningitis, iv, 738  
 cervical, iv, 733  
 in chancroid, iv, 739  
 in chickenpox, iv, 737  
 chronic, iv, 732  
 in diphtheria, iv, 736  
 in erysipelas, iv, 737  
 of femoral glands, iv, 736  
 in foot and mouth disease, iv, 738  
 in glanders, iv, 737  
 in glandular fever, iv, 738
- Lymphadenitis in infectious jaundice, iv, 738  
 in influenza, iv, 737  
 of inguinal glands, iv, 736  
 in malarial fever, iv, 738  
 in Malta fever, iv, 738  
 in measles, iv, 736  
 of mesenteric glands, iv, 736  
 in miliary fever, iv, 738  
 in mumps, iv, 737  
 in pneumococcus septicemia, iv, 738  
 purulenta, iv, 731  
 in pyemia, iv, 737  
 in relapsing fever, iv, 738  
 in rheumatic fever, i, 999; iv, 737  
 in Rocky Mountain spotted fever, iv, 738  
 in rubella, iv, 736  
 in scarlet fever, iv, 736  
 in septicemia, iv, 737  
 simplex, iv, 730  
 in smallpox, iv, 737  
 in syphilis, iv, 740  
 in trypanosomiasis, iv, 738  
 tuberculous, iv, 741  
 in typhoid fever, iv, 738  
 in typhus fever, iv, 738  
 in vaccinia, iv, 737
- Lymphadenoma of thymus, iv, 932  
 Lymphangiectasia, iv, 596  
 Lymphangio-endothelioma, iv, 600  
 Lymphangioma, iv, 596  
 of intestines, iii, 434  
 of lips, iii, 45  
 Lymphangitis, iv, 592  
 filarial, ii, 308  
 productiva, iv, 595  
 Lymphangivitis of thoracic duct, iv, 584  
 Lymphatic channels, tuberculosis of, i, 359  
 circulation, iv, 752  
 biliary infection and, iii, 551  
 system in exophthalmic goitre, iv, 856  
 Lymphocytes in leukemia, iv, 667, 674  
 Lymphocytoma, iv, 748  
 Lymphocytosis, iv, 615  
 in exophthalmic goitre, iv, 867  
 in spinal fluid, v, 177  
 Lymphogenous lymphadenitis, iv, 729  
 Lymphoid leukemia, iv, 670  
 tissue, appendicitis and, iii, 361  
 tuberculosis of lymph nodes, iv, 741  
 Lymphoma of pericardium, iv, 76  
 of thymus, iv, 932  
 Lymphomata, iv, 748  
 Lymphorrhagia, iv, 595  
 in elephantiasis, iv, 598  
 in myasthenia gravis, v, 799  
 Lymphosarcoma of intestines, iii, 431  
 of lymphatic vessels, iv, 601  
 of mediastinum, ii, 1094  
 of thymus, iv, 932
- Lysins, i, 54

## M

- McBURNAY's point in appendicitis, iii, 367  
 Macewen's sign in cerebrospinal meningitis, i, 608

- Macrocephaly in achondroplasia, v, 963  
 Macrogamete, ii, 61  
 Macrogametocytes, ii, 61  
 Macroglossia, iii, 60  
 Madura foot, i, 1049  
 Magnesium phosphate calculus, iii, 1015  
 Mal des montagnes, ii, 344  
 Malarial fevers, ii, 54  
     acute endocarditis and, iv, 157  
         hepatitis and, iii, 494  
     anemia and, iv, 658  
     angioneurotic edema and, iv, 999  
     cirrhosis of liver and, iii, 509, 530  
     estivo-autumnal, quotidian, ii, 80  
     inflammation of spleen and, iv, 941  
     irregular, ii, 84  
     latent, ii, 85  
     masked, ii, 85  
     migraine and, v, 720  
     nephritis and, iii, 850  
     pernicious, ii, 80  
         anemia and, iv, 622  
         pigmentation of liver and, iii, 470  
         quartan, ii, 77  
         remittent, ii, 84  
         tertian, ii, 74  
         tuberculosis and, i, 322  
         typhoid fever and, i, 156; ii, 87  
 Malformations of kidney, iii, 785  
     of spleen, iv, 937  
 Malignant edema, i, 645  
     pustule, i, 639, 644  
 Mallein, i, 650  
 Malnutrition, ii, 666  
     kidney infections and, iii, 952  
     multiple neuritis and, v, 576  
 Malpighian bodies in nephritis, iii, 858, 896  
 Malpositions of spleen, iv, 938  
 Malta fever, i, 644  
 Mammary glands in exophthalmic goitre, iv, 868  
     -in tuberculosis, i, 435  
     neuralgia, v, 739  
 Manchurian "typhus," i, 199  
 Manganese poisoning, ii, 386  
 Mania, acute, traumatism and, v, 647  
     in lead poisoning, ii, 368  
     in uremia, iii, 837  
 Marie's disease, i, 616  
     hereditary cerebellar ataxia, v, 130, 136  
     theories on aphasia, v, 300  
 Mast-cells, iv, 616  
     in myeloid leukemia, iv, 668  
 Mastitis in rheumatic fever, i, 999  
     in typhoid fever, i, 145  
 Mastodynia, v, 739  
 Mastoiditis in scarlet fever, i, 875  
 Measles, i, 897  
     apoplexy and, v, 417  
     arteriosclerosis and, iv, 454  
     chickenpox and, i, 907  
     malignant, i, 903  
     nephritis and, iii, 849  
     pregnancy and, i, 907  
     smallpox and, i, 907  
     tuberculosis and, i, 321, 907  
 Measles, whooping cough and, i, 659  
 Meat poisoning, ii, 458  
 Meckel's diverticulum, iii, 421  
 Mediastinal emphysema, ii, 1110  
     hemorrhage, ii, 1110  
     hernia, ii, 1109  
 Mediastinitis, ii, 1104  
     in lobar pneumonia, i, 253  
     pericarditis and, iv, 59, 62  
 Mediastinum, carcinoma of, ii, 1100  
     cysts of, ii, 1101, 1109  
     endothelioma of, ii, 1100  
     lymph nodes of, hyperplasia of, ii, 1110  
         pigmentation of, ii, 1111  
         sclerosis of, ii, 1111  
         tuberculosis of, ii, 1111  
     sarcoma of, ii, 1094  
     teratoma of, ii, 1101  
     tumors of, ii, 1093  
 Mediterranean fever, i, 664  
 Medulla oblongata, foraminal hernia of, v, 324  
     tumors of, v, 337  
 Megaloblasts in pernicious anemia, iv, 638  
 Megalocephalia, v, 988  
 Megalopsia, v, 480  
 Megastoma entericum in carcinoma of stomach, iii, 213  
 Megrims, v, 709  
 Melancholia in pernicious anemia, iv, 631  
 Melanemia in malarial fevers, ii, 68  
 Melanoglossia, iii, 61  
 Melanuria, iii, 811  
 Membranous cholecystitis, iii, 563  
     colitis, iii, 437  
 Memory, anomalies of, v, 53, 55  
 Mendel's sign, v, 78  
 Ménière's disease, v, 512  
     syndrome in brain tumor, v, 337  
 Meninges in cerebrospinal syphilis, v, 824  
     diseases of, v, 162  
 Meningism in lobar pneumonia, i, 260  
     in typhoid fever, i, 137  
 Meningitis, acute cerebrospinal, v, 167  
     epidemic cerebrospinal, i, 594  
     epilepsy and, v, 599  
     in influenza, i, 537, 543  
     in lobar pneumonia, i, 260  
     localized serous spinal, v, 174  
     posterior basic, i, 615  
         hydrocephalus and, v, 357  
     in scarlet fever, i, 878  
     serous, ii, 415  
         hydrocephalus and, v, 358  
         tuberculous, i, 420, 434  
         cardiac arrhythmia and, iv, 287  
         hydrocephalus and, v, 357  
     in typhoid fever, i, 137  
 Meningo-encephalitis, tuberculous, of brain, v, 319  
 Meningomyelitis, gonococcal, i, 761  
     in syphilis, ii, 162  
 Mental disturbances, traumatic, v, 646  
     fog, v, 44  
     phenomena in tetany, v, 758  
 Meralgia paresthetica, v, 557  
 Mercurial embolism, iv, 561

- Mercurial stomatitis, iii, 48  
 Mercury poisoning, ii, 378  
     purpura and, iv, 689  
     in syphilis, ii, 207  
 Mermithidae, ii, 296  
 Merozoites, ii, 58, 59, 60  
 Merycism, iii, 138  
 Mesaortitis, acute, iv, 451  
 Mesenteric cysts of peritoneum, iii, 736  
     embolism, iii, 434  
     glands, acute peritonitis and, iii, 673  
     lymphadenitis of, iv, 736  
     tuberculosis of, iv, 743  
     in typhoid fever, i, 85  
     thrombosis, iii, 434; iv, 542  
     cirrhosis of liver and, iii, 435  
 Metabolism, ii, 549  
     in acromegaly, iv, 812  
     in arthritis deformans, v, 903, 907  
     auto-intoxication and, ii, 505  
     of calcium, ii, 561, 636  
     carbohydrate, ii, 596, 687  
     of carbon, ii, 560  
     cholelithiasis and, iii, 571  
     in diabetes mellitus, ii, 543, 686, 691  
     during fasting, ii, 606  
     energy, ii, 564  
     in exophthalmic goitre, iv, 869  
     fat, ii, 601  
     in gout, ii, 732  
     of hexosephosphate, ii, 562  
     in hypopituitarism, iv, 816  
     in hysteria, v, 688  
     in infections, i, 44  
     of lecithins, ii, 562  
     in lobar pneumonia, i, 217  
     of magnesium, ii, 561  
     multiple neuritis and, v, 575  
     in myxoedema, ii, 517  
     nephritis and, iii, 854, 892  
     of nitrogen, ii, 556  
     of nucleic acid, ii, 562  
     in obesity, ii, 516, 666  
     osteomalacia and, v, 949, 952  
     of phosphoproteins, ii, 562  
     of phosphorus, ii, 561, 637  
     protein, ii, 579, 582  
     purpura and, iv, 700  
     spleen and, iv, 934  
     in sporadic cretinism, iv, 885  
     of sulphur, ii, 561  
     in tuberculosis, i, 381  
     of uric acid, ii, 732  
 Metacarpophalangeal joints in arthritis deformans, v, 922  
 Metallic tinkle in pneumothorax, ii, 1080  
 Metapneumonic pleuritis, ii, 1012  
 Metastasis in mumps, i, 921  
 Metastatic abscess of brain, v, 384  
 Metastrongylus apisi, ii, 302  
 Metasyphilis, ii, 185  
 Meteorism, iii, 298  
     in angioneurotic edema, iv, 1008  
     in hepatoptosis, iii, 757  
     in lobar pneumonia, i, 254  
     in typhoid fever, i, 106  
 Methyl alcohol poisoning, ii, 398  
 Meyer's sensitized bacillus emulsions, i, 305  
 Microcephalia, v, 990  
 Micrococcus of endocarditis, i, 559  
     gingivae in mouth, iii, 40  
     melitensis, i, 664  
     paramelitensis, i, 665  
     rheumaticus, i, 976  
 Microfilaria, ii, 305, 315  
 Microgametes, ii, 61  
 Microgametocyte, ii, 61  
 Microglossia, iii, 60  
 Micromelic rickets, v, 955  
 Mid-brain, glioma of, v, 284  
     lesion of, v, 282  
 Migraine, epilepsy and, v, 615  
     ophthalmic, v, 709  
     gout and, ii, 748; v, 720  
     ophthalmoplegic, v, 724  
     periodic paralysis and, v, 811  
 Mikulicz's disease, iii, 73  
 Miliary aneurisms, hematemeses from, iii, 283  
     eruption in typhoid fever, i, 101  
     fever, i, 1017  
     sclerosis, v, 161  
     tuberculosis, acute, i, 417  
 Milk poisoning, ii, 466  
     sickness, i, 1028  
 Milky ascites, iii, 662  
 Millard-Gubler paralysis, v, 290, 369  
 Milroy's disease, iv, 1011  
 Mind blindness, v, 53, 271, 299, 483  
     deafness, v, 54  
 Miner's anemia, ii, 271  
     cachexia, ii, 271  
 Miosis, v, 77  
     in exophthalmic goitre, iv, 864  
 Mitral atresia, congenital, iv, 406  
     insufficiency, iv, 232  
         in tuberculosis, i, 433  
     stenosis, iv, 239  
         in tuberculosis, i, 433  
     valve, division of, iv, 359  
 Moebius sign in exophthalmic goitre, iv, 863  
 Moeller's glossitis, iii, 63  
 Mollities ossium, v, 947  
 Molluscum fibrosum, v, 571  
 Monarticular arthritis deformans, v, 919, 934  
 Monocular diplopia, v, 480  
 Monoideic somnambulism, v, 44  
 Monoplegia in arteriosclerosis, iv, 461  
     in hysteria, v, 681  
     in tuberculosis, i, 421  
 Monostomulum lentis, ii, 245  
 Moods, v, 59, 62  
 Morbus caeruleus, iv, 333  
     gallicus, ii, 144  
     herculeus, iv, 596  
 Morphimism, ii, 438  
 Morvan's type of syringomyelia, v, 220  
 Mosquito brigades, ii, 97  
     malarial, ii, 63  
 Motility, disturbances of, v, 68  
 Motor actions, v, 64, 65, 66

- Motor agitation, v, 63  
 aphasia, v, 75, 292, 295  
 area of cortex of brain, v, 265  
 disturbances in migraine, v, 715  
   in neuralgia, v, 728  
 functions of intestines, iii, 286  
 insufficiency of stomach, iii, 247  
 nerves in tetany, v, 757  
 paralysis of intestines, iii, 418  
 phenomena in infantile convulsions,  
 preservation, v, 67  
   v, 794  
 stupor, v, 63, 65
- Mountain sickness, ii, 344
- Mouth, acne of, iii, 54  
 actinomycosis of, iii, 59  
 angioneurotic oedema of, iii, 53  
 carcinoma of, iii, 52  
 cysts of, iii, 52  
 dermatoses of, iii, 53  
 in diabetes, iii, 39  
 eczema of, iii, 54  
 erysipelas of, iii, 54  
 in foot-and-mouth disease, i, 1027  
 in hemiplegia, v, 460  
 inflammation of, iii, 45  
 leprosy of, iii, 59  
 in leukemia, iii, 55  
 lichen planus of, iii, 54  
 in locomotor ataxia, iii, 55  
 lupus erythematosus of, iii, 53  
 pemphigus of, iii, 54  
 in pernicious anemia, iii, 55; iv, 630  
 in pregnancy, iii, 56  
 psoriasis of, iii, 53  
 scleroderma of, iii, 54  
 scleroma of, iii, 60  
 in scurvy, ii, 800; iii, 55  
 sepsis of, iii, 41  
 in sprue, iii, 393  
 syphilis of, iii, 58  
 tinea circinata of, iii, 54  
 tuberculosis of, i, 360; iii, 57  
 tumors of, iii, 52  
 urticaria of, iii, 53  
 in varicella, ii, 56  
 in variola, iii, 56
- Movement, sensation of, v, 17
- Mucocoele of appendix, iii, 386
- Mucous colic, iii, 437  
 colitis, iii, 437  
 diarrhoea, iii, 437  
 gastritis, iii, 238  
 lesions in syphilis, ii, 163  
 membrane in acute laryngitis, ii, 869  
   in angioneurotic oedema, iv, 1007  
   in asthma, ii, 932  
   endocarditis and, iv, 152  
   in hay fever, ii, 855, 857, 864  
   in hysteria, v, 682  
   in jaundice, iii, 451  
   in pellagra, ii, 479, 480  
   in syphilis, ii, 151, 168  
     of larynx, ii, 872  
   in tuberculosis of larynx, ii, 876  
   patches in congenital syphilis, ii, 189
- Mucus in feces, iii, 293
- Mulberry calculus, iii, 1009
- Multiple neuritis, v, 593  
 sclerosis, v, 140
- Mumps, i, 918
- Mural aneurism, iv, 483
- Murmur, cardiopulmonary, iv, 298
- Muscles, abdominal, constipation and, iii,  
 312  
   in acromegaly, iv, 811  
   in acute poliomyelitis, i, 736  
   in amyotonia congenita, v, 894  
   in arthritis deformans, v, 907  
   atony of, v, 68  
   atrophy of, v, 68  
   in disease of peripheral nerves, v,  
   534  
   in paralysis agitans, v, 771  
   contractures of, v, 68  
   cramps of, in arteriosclerosis, iv, 464  
   in tetany, v, 754  
   electrical reactions of, v, 72  
   in exophthalmic goitre, iv, 857, 865  
   hypertony of, v, 68  
   hypertrophy of, v, 68  
   hypotony of, v, 68  
   in tabes dorsalis, v, 839  
   inflammation of, v, 871  
   irritation of, v, 70  
   in myositis ossificans, v, 883  
   in myotonia congenita, v, 887  
   of neck, spasm of, v, 525  
   ocular, paralysis of, v, 485  
   paralysis of, v, 68  
   in paramyoclonus multiplex, v, 807  
   in periodic paralysis, v, 813  
   in rickets, ii, 780  
   in scleroderma, iv, 1020  
   sense, v, 17  
   spasms of, v, 71  
   of tongue, spasm of, v, 532  
   tonus of, v, 68  
   tuberculosis of, i, 373
- Muscular atrophy, v, 80  
   in arthritis deformans, v, 914  
   progressive spinal, v, 82  
 dystrophy, progressive, v, 117  
 rigidity in hemiplegia, v, 463  
   in paralysis agitans, v, 769  
 tension in hemiplegia, v, 463  
 wasting in spondylitis, v, 927  
 weakness in chlorosis, iv, 649  
   in general paresis, v, 851  
   in pernicious anemia, iv, 629  
   in rickets, ii, 782
- Musculocutaneous nerve, paralysis of, v,  
 546
- Musculospiral nerve, paralysis of, v, 547
- Mutism in hysteria, v, 652
- Myasthenia, dilatation of stomach and, iii,  
 255  
   gravis, v, 798  
   in hysteria, v, 678  
   parathyroid glands and, iv, 903
- Myasthenic reaction, v, 801, 805
- Mycetoma, i, 1049
- Mycobacterium tuberculosis, i, 294
- Mycoses, i, 1032

- Mycosis intestinalis, i, 645  
   mucorina, i, 1060  
   of tonsils, ii, 852  
 Mycotic aneurism, iv, 476  
   gastritis, iii, 235  
   stomatitis, iii, 235  
 Mydriasis v, 77  
   in exophthalmic goitre, iv, 864  
   in lobar pneumonia, i, 263  
   in tabes dorsalis, v, 837  
 Myelitis, v, 227  
   acute ascending, v, 236  
     bulbar, v, 368  
   chronic, v, 245  
   compression, v, 229  
   degenerative, v, 236  
   disseminated, v, 235  
   infective, v, 229  
   syphilitic, v, 237, 828  
   toxic, v, 236  
 Myelocytes in lobar pneumonia, i, 234  
   in myeloid leukemia, iv, 667  
   in pernicious anemia, iv, 638  
 Myelocytosis, iv, 616  
 Myeloid leukemia, iv, 661  
 Myeloma of brain, v, 315  
   of spinal cord, v, 198  
 Myelophthisic anemia, iv, 645  
 Myeloplastic bone aplasia, v, 978  
 Myiasis, ii, 328  
 Myocardial bradycardia, iv, 282  
   incompetence of obese, iv, 128  
 Myocarditis, iv, 108  
   acute interstitial, iv, 113  
     necrotic, iv, 114  
     parenchymatous, iv, 113  
   angina pectoris and, iv, 291  
   in arteriosclerosis, iv, 462  
   cardiac arrhythmia and, iv, 287  
   chronic, iv, 109  
   in endocarditis, iv, 161  
   gonorrhœal, i, 752  
   in gout, ii, 749  
   hypertrophy of heart and, iv, 182  
   in measles, i, 907  
   in pericarditis, iv, 45  
   in scarlet fever, i, 877  
   in smallpox, i, 811  
   syphilitic, ii, 184  
   tricuspid insufficiency and, iv, 247  
   in typhoid fever, i, 126  
   in typhus fever, i, 930  
 Myocardium, actinomycosis of, iv, 145  
   blastomycosis of, iv, 144  
   degeneration of, iv, 108-116  
   in diphtheria, i, 694  
   echinococcus of, iv, 147  
   fragmentation of, iv, 116  
   insufficiency of, iv, 131, 187  
   ischemia of, iv, 290  
   new growths of, iv, 142, 145  
   parasites of, iv, 142  
   segmentation of, iv, 116  
   syphilis of, iv, 142  
   tuberculosis of, i, 359; iv, 144  
 Myoclonus in hysteria, v, 677  
   parathyroid glands and, iv, 903  
 Myogenic theory of mechanism of heart, iv, 27  
 Myoidema in tuberculosis, i, 411  
 Myoma of appendix, iii, 386  
   of peritoneum, iii, 730  
 Myopathy, primary, v, 117  
 Myopia, v, 36  
 Myositis, v, 871  
   gonococcal, i, 760  
   non-suppurative, i, 760; v, 875  
   ossificans, progressive, v, 881  
   primary fibrosa, v, 880  
     suppurative, i, 761; v, 872  
   with special terminal lesions, v, 880  
 Myospasm caused by heat exposure, ii, 351  
 Myotonia congenita, v, 886  
   parathyroid glands and, iv, 903  
 Myotonic reaction, v, 891  
 Myriapoda, ii, 324  
 Mythomania, v, 659  
 Mytilotoxin, ii, 457  
 Myxœdema, iv, 888  
   congenital, iv, 883  
   in exophthalmic goitre, iv, 869, 890  
   goitre and, iv, 890  
   infantile, iv, 883  
   spontaneous, iv, 888  
   thyroiditis and, iv, 829  
 Myxoma of appendix, iii, 386  
   of intestines, iii, 433  
   of peritoneum, iii, 733  
 Myxosarcoma of brain, v, 315

## N

- NAGANA, ii, 117  
 Nails in diseases of peripheral nerves, v, 541  
   in endemic cretinism, iv, 882  
   in osteo-arthritis, v, 984  
   in syphilis, ii, 167  
   in typhoid fever, i, 102  
 Nasal diphtheria, i, 698  
 Nasopharyngitis, ii, 837, 838  
 Nasopharynx, adenoid growths in, ii, 840  
   tuberculosis of, i, 389  
 Nauheim baths in myocarditis, iv, 138  
 Necator americanus, ii, 272  
 Neck, actinomycosis of, i, 1038  
   in cerebrospinal meningitis, i, 607  
   muscles of, spasm of, v, 525  
 Necrophagus sanguinarius, ii, 323  
 Necrosis of appendix, iii, 363  
   fat, iii, 606  
   hemorrhagic, of pancreas, iii, 610  
   of jaw in phosphorus poisoning, ii, 383  
   of liver, iii, 471  
   in Raynaud's disease, iv, 986  
 Necrotic endocarditis, iv, 151  
 Negri bodies, i, 959, 961  
 Nemathelminthes, ii, 270  
 Nematodes of urogenital system, ii, 315  
 Neoplasms of lymph nodes, iv, 747, 752  
   of lymphatic vessels, iv, 600, 601  
   of pericardium, iv, 74  
 Neosalvarsan in syphilis, ii, 212



- Nephritis, iii, 778, 845  
 acute, iii, 857  
   alcoholism and, iii, 866  
   in Asiatic cholera, iii, 867  
   in chronic nephritis, iii, 912  
   in Henoch's purpura, iv, 710  
   primary, iii, 867  
   in purpura hemorrhagica, iv, 712  
   in typhoid fever, iii, 867  
   yellow atrophy of liver and, iii, 850  
 arteriosclerosis and, iv, 455  
 burns and, iii, 853  
 in chickenpox, i, 853  
 cholera and, iii, 849  
 chorea and, iii, 850  
 chronic, diffuse, iii, 847  
   interstitial, iii, 890  
   myocarditis and, iv, 123, 125  
   in myxœdema, iv, 894  
   parenchymatous, iii, 880  
   pericarditis and, iv, 42  
   pernicious anemia and, iv, 627  
 diarrhœa and, iii, 323  
 in diphtheria, i, 695; iii, 849  
 in glandular fever, i, 1013  
 lobar pneumonia and, i, 208, 258  
 in malarial fevers, ii, 86; iii, 850  
 in mumps, i, 921  
 in polymyositis hemorrhagica, v, 880  
 in rheumatic fever, i, 998; iii, 850  
 in scarlet fever, i, 874; iii, 849  
 in serofibrinous pleuritis, ii, 1026  
 in smallpox, i, 812; iii, 849  
 in syphilis, ii, 180  
 tuberculosis and, i, 434  
 in typhoid fever, i, 143; iii, 849
- Nephrolins, iii, 834
- Nephroptosis, iii, 750, 755  
 kidney infections and, iii, 952  
 tuberculosis of kidney and, iii, 978
- Nerve deafness, v, 509  
 trunks, neuralgia and, v, 726
- Nerves in acute poliomyelitis, i, 736  
 in beriberi, ii, 493  
 in cerebrospinal meningitis, i, 598  
 in goitre, iv, 842  
 tumors of, v, 568
- Nervous anorexia, iii, 157  
 dyspepsia, iii, 164  
 dyspnœa, iv, 37  
 eructation, iii, 141  
 gastralgia, iii, 135  
 hypertrophy of heart, iv, 167  
 influenza, i, 542  
 nausea, iii, 146  
 purpura, iv, 690  
 shock, pernicious anemia and, iv, 624  
 system in Addison's disease, iv, 793  
   in alcoholism, ii, 405  
   angioneurotic œdema and, iv, 999  
   in arteriosclerosis, iv, 461  
   arthritis deformans and, v, 903, 907  
   in cardiac insufficiency, iv, 208  
   in chlorosis, iv, 650  
   in dermatomyositis, v, 877
- Nervous system, diabetes mellitus and, ii, 678, 697  
 in diphtheria, i, 695  
 in exophthalmic goitre, iv, 857, 864  
 in jaundice, iii, 454  
 in lead poisoning, ii, 365  
 in lobar pneumonia, i, 260  
 in measles, i, 907  
 in motor insufficiency of stomach, iii, 248  
 in myxœdema, iv, 892  
 in opium smoking, ii, 436  
 in Paget's disease, v, 973  
 parasymphilis of, v, 822  
 in pellagra, ii, 477, 481  
 in pernicious anemia, iv, 628  
 in plague, i, 630  
 in purpura, iv, 697  
 in rheumatic fever, i, 992  
 syphilis of, v, 822  
 in tuberculosis, i, 390, 421  
 in typhoid fever, i, 80, 132  
 in uremia, iii, 835  
 vomiting, iii, 142
- Neuralgia, v, 725  
 brachial, v, 737  
 cervico-occipital, v, 736  
 in chronic nephritis, iii, 907  
 of coccygeal plexus, v, 741  
 diaphragmatic, v, 737  
 facial, v, 733  
 femoral, v, 740  
 ganglia and, v, 726  
 gonococcal, i, 761  
 in gout, ii, 748  
 in hemophilia, iv, 725  
 in hysteria, v, 683  
 intercostal, v, 739  
 of intestines, iii, 305  
 of lumbar plexus, v, 739  
 lumbo-abdominal, v, 739  
 mammary, v, 739  
 nerve trunks and, v, 740  
 obturator, v, 740  
 of pudendal plexus, v, 740  
 "red painful," iv, 1028  
 reflex, v, 726, 731  
 testicular, v, 740  
 trigeminal, v, 733  
 Valleix's points in, v, 727
- Neurasthenia, v, 624  
 achylia gastrica and, iii, 153  
 in arteriosclerosis of brain, v, 395  
 cardiospasm and, iii, 147  
 in constipation, iii, 315  
 electrical shock and, ii, 341  
 gastric, iii, 164  
 in mucous colitis, iii, 439  
 nervous anorexia and, iii, 157  
   dyspepsia and, iii, 165  
 in splanchnoptosis, iii, 753, 754  
 syphilis and, v, 629  
 in tabes dorsalis, v, 843  
 traumatic, v, 643
- Neuritic pains in arteriosclerosis, iv, 465
- Neuritis in arsenic poisoning, ii, 375

- Neuritis, brachial, v, 553  
     in diabetes mellitus, ii, 706, 708  
     gonococcal, i, 761  
     in hemophilia, iv, 725  
     in influenza, i, 543  
     interstitial hypertrophic progressive,  
         of childhood, v, 112  
     in lobar pneumonia, i, 262  
     in mercury poisoning, ii, 380  
     multiple, v, 573  
         alcoholic, v, 579  
         ascending, v, 576  
     optic, v, 36  
         in acute meningitis, v, 170  
     retrobulbar, v, 37, 479  
     in rheumatic fever, i, 995  
     in tuberculosis, i, 392  
     in typhoid fever, i, 139  
 Neuroblastoma of brain, v, 317  
 Neurocytes hydrophobia, i, 959  
 Neurocytoma of brain, v, 317  
 Neurofibromatosis, general, v, 570  
 Neuroglia in infective myelitis, v, 232  
     in syphilitic myelitis, v, 239  
 Neuroglioma of brain, v, 311  
 Neuroma of brain, v, 317  
     gangliated, of peritoneum, iii, 731  
 Neuromata, v, 568  
 Neuromyositis, v, 880  
 Neuropathic hematemeses, iii, 282  
 Neuroses, cardiovascular disease and, iv,  
     296  
     gastric, iii, 106  
     gonococcal, i, 762  
     of larynx, sensory, v, 522  
     occupation, v, 741  
     of cesophagus, iii, 86  
     polysymptomatic, iii, 164  
     professional, v, 741  
     traumatic, v, 642  
     in tuberculosis, i, 391  
 Neuro-tabes peripherica, v, 579, 584  
 Neurotic progressive facial atrophy, v, 991  
 Neutrophiles in myeloid leukemia, iv, 667  
 Neutrophilic myelocytes, iv, 609  
 Night-blindness, v, 35, 480  
     in scurvy, ii, 802  
 Night-sweats in tuberculosis, i, 381  
 Nitric oxide poisoning, ii, 395  
 Nitrogen balance, ii, 556  
     equilibrium, ii, 558  
     in feces, ii, 640  
     metabolism of, ii, 556  
     non-protein, in metabolism, ii, 662  
     in urine in diabetes mellitus, ii, 703  
 Nocardiosis of pleura, ii, 1051  
 Nodosity of joints, v, 897  
 Nodules in septic endocarditis, iv, 163  
 Noma, iii, 49  
 Normoblasts in pernicious anemia, iv, 638  
 Nose in cerebrospinal meningitis, i, 604  
     in glanders, i, 652  
     obstruction of, ii, 815  
     tuberculosis of, i, 347  
     in typhoid fever, i, 120, 140  
 Nyctalopia, v, 35, 480  
 Nystagmus, v, 70  
     Nystagmus in brain abscess, v, 384  
         tumor, v, 336, 339, 340  
     in disseminated sclerosis, v, 151  
     in Friedreich's ataxia, v, 135  
     in lead poisoning, ii, 368  
     in Ménière's disease, v, 513  
     in mercury poisoning, ii, 379  
     in ocular paralysis, v, 486  
     in pseudo-sclerosis, v, 159  
     in syringomyelia, v, 222  
     in vestibular disease, v, 511
- O**
- OBESITY, ii, 761  
     fat metabolism and, ii, 605  
     myocardial incompetence of, iv, 128  
 Oblique muscles, paralysis of, v, 485  
 Obliterative tuberculosis of peritoneum, iii,  
     724  
 Obsessions, v, 52  
 "Obstetrical hand," v, 71  
 Obstipation, iii, 309  
 Obstruction of intestines, iii, 394  
 Obturation of intestines, iii, 394  
 Obturator nerve, paralysis of, 557  
 Occlusion of intestines, iii, 394  
 "Occult blood" in carcinoma of stomach,  
     iii, 207, 211  
     in gastric ulcer, iii, 179  
     tests for, iii, 180  
 Occupation neuroses, v, 741  
 Ochronosis, iii, 817  
 Ocular muscles in disseminated sclerosis,  
     v, 151  
     paralysis of, v, 485  
     nerves, paralysis of, 483  
     palsy, nuclear, v, 365  
 Oculomotor palsy, recurrent, v, 488  
     in tabes dorsalis, v, 841  
 O'Dwyer's intubation instruments, i, 726  
 Edema, angioneurotic, iv, 998  
     of mouth, iii, 53  
     of pharynx, ii, 847  
     of larynx, ii, 878  
     of legs, hereditary, iv, 1011  
     of lungs, ii, 948  
 Esophageal nerves, paralysis of, v, 523  
 Esophagitis, iii, 79  
 Esophagostomum, ii, 294  
 Esophagus, actinomycosis of, iii, 86  
     in aneurism of aorta, iv, 502, 509  
     carcinoma of, iii, 98  
     congenital malformations of, iii, 78  
     dilatation of, iii, 90  
     diverticula of, iii, 92  
     in goitre, iv, 843  
     herpes of, iii, 90  
     inflammation of, iii, 79  
     neuroses of, iii, 86  
     non-malignant growths of, iii, 105  
     paralysis of, iii, 90  
     perforation of, ii, 1045  
     rupture of, iii, 85  
     sarcoma of, iii, 105  
     stenosis of, iii, 96

- Œsophagus, stricture of, iii, 96  
   syphilis of, ii, 174; iii, 86  
   tuberculosis of, i, 361; iii, 85  
   in typhoid fever, i, 104  
   ulcers of, iii, 84  
 Oïdiomycosis, i, 1050  
 Oidium albicans, i, 1054; iii, 46, 50  
 Olfactory nerve, diseases of, v, 471  
 Oliguria in tuberculosis of kidney, iii, 982  
 Olivopontocerebellar atrophy, v, 287  
 Omental cysts of peritoneum, iii, 737  
 Omentitis, mesenteric lymphadenitis and, iv, 736  
 Omentum, abscess of, iii, 696  
   inflammation of, iii, 695  
   volvulus of, iii, 418  
 Oöcyst, ii, 63  
 Oökinete, ii, 62  
 Ophthalmic distomatosis, ii, 245  
 Ophthalmoplegia, v, 483  
   in acute encephalomyelitis, v, 369  
   in cerebrospinal syphilis, v, 825  
   chronic, v, 491  
   externa, v, 490  
   in hysteria, v, 678  
   interna, v, 490  
   in lesion of quadrigeminal bodies, v, 283  
   in superior polienccephalitis, v, 365  
   in tabes dorsalis, v, 841  
 Ophthalmoplegic migraine, 724  
 Ophthalmic-reaction in typhoid fever, i, 162  
 Opisthorchiasis, ii, 225, 232, 233  
 Opisthorchis, ii, 229, 232  
 Opium habit, ii, 433  
   poisoning, ii, 433  
   smoking, ii, 435  
 Oppenheim's disease, v, 893  
   sign, v, 78  
 Opsonic index in septicemia, i, 569, 573  
   in tuberculosis, i, 511  
 Opsonins, i, 57  
   in tuberculosis, i, 313  
 Optic aphasia, v, 54, 299  
   asymbolia, v, 54  
   atrophy, v, 478  
   chiasm, affections of, v, 481  
   disks in disseminated sclerosis, v, 151  
   in papillitis, v, 478  
   nerve, atrophy of, v, 37  
   in tabes dorsalis, v, 841  
   inflammation of, v, 476  
   neuritis, v, 36, 326  
   in diabetes mellitus, ii, 708  
   in tuberculosis, i, 421  
   thalamus, lesion of, v, 279  
   tracts, affections of, v, 481  
 Oral sepsis, iii, 41  
 Orchitis in congenital syphilis, ii, 190  
   in filariasis, ii, 310  
   in lobar pneumonia, i, 264  
   in mumps, i, 921  
   in typhoid fever, i, 145  
 Oriental sore, iv, 964  
 Ornithodoros, ii, 317  
 Orthopnoea in cardiac insufficiency, iv, 296  
 Osteitis, chronic parenchymatous, v, 978  
 Osteitis deformans, v, 969  
   tumor-building, v, 978  
   fibrosa, 980  
   in syphilis, ii, 163  
 Osteo-arthritis, v, 897, 920  
   hypertrophica, v, 987  
 Osteo-arthropathy, hypertrophic, v, 981  
   in tuberculosis, i, 411  
 Osteochondritis in syphilis, ii, 190  
 Osteogenesis imperfecta, v, 907  
 Osteoma of lung, ii, 997  
 Osteomalacia, v, 947  
   congenita, v, 978  
   in exophthalmic goitre, iv, 868  
   parathyroid glands and, iv, 903  
 Osteopathies, hypertrophic, v, 969  
 Osteoporosis congenita, v, 978  
 Osteopsathyrosis, v, 978, 980  
 Osteosarcoma of brain, v, 314  
 Osteosclerosis, congenital, v, 968, 978  
 Otitis deformans, v, 970  
   fibrosa hypertrophica, v, 988  
 Ostium primum, iv, 326  
   secundum, iv, 326  
 Otitic abscess of brain, v, 383  
 Otitis media in chickenpox, i, 853  
   in diphtheria, i, 700  
   in glandular fever, i, 1013  
   in influenza, i, 537, 545  
   in lobar pneumonia, i, 263  
   in scarlet fever, i, 864, 875  
   in smallpox, i, 812  
   in typhoid fever, i, 140  
 Ovarian cyst, acute peritonitis and, iii, 673  
 Ovaries in osteomalacia, v, 948  
   tuberculosis of, i, 368  
   in typhoid fever, i, 144  
 Ovaritis in mumps, i, 921  
 Oxalic acid calculus, iii, 1015  
 Oxaluria, ii, 540; iii, 815  
   in diabetes mellitus, ii, 703  
   in gout, ii, 748  
 Oxyuriasis, ii, 291  
 Oxyuris vermicularis, ii, 291  
   appendicitis and, iii, 360

## P

- PACHYDERMA, iv, 596  
 Pachymeningitis, v, 163  
   cervicalis hypertrophica, v, 166  
   externa, v, 163  
   interna, v, 164  
 Paget's disease, v, 970  
 Pain sense, v, 17  
 Painful foot, i, 760  
   heel, i, 760  
 Palate in hemiplegia, v, 459  
   paralysis of, in hysteria, v, 679  
   soft, paralysis of, iii, 62  
   tuberculosis of, iii, 57  
 Pallanesthesia, v, 30  
 Palmar reflex, v, 78  
 Palsy, bulbar, acute inflammatory, v, 368  
   progressive, v, 113  
   congenital cerebral, v, 410

- Palsy, Duchenne-Erb, v, 551  
     Klumpke's, v, 552  
     oculomotor, recurrent, v, 488  
         in tabes dorsalis, v, 841  
     shaking, v, 764  
 Panatrophphy, local, in scleroderma, iv, 1022  
 Pancreas, abscess of, calculus and, iii, 635  
     subphrenic abscess and, iii, 701  
     adenoma of, iii, 649  
     arteriosclerosis of, iii, 605  
     calculus of, iii, 632  
         carcinoma and, iii, 636  
     carcinoma of, iii, 648  
         calculus and, iii, 636  
     cyst of, iii, 637  
         diabetes mellitus and, iii, 645  
         necrosis and, iii, 619  
     descent of, iii, 758  
     dilatation of stomach and, iii, 251  
     diseases of, diabetes mellitus and, iii, 604  
         glycosuria and, iii, 604, 606  
         metabolic function and, iii, 604  
         pancreatic secretion and, iii, 599  
     displaced, iii, 752  
     fat necrosis and, iii, 606  
     gangrene of, iii, 615  
     hemorrhagic necrosis of, iii, 610  
     in hypopituitarism, iv, 817  
     sarcoma of, iii, 650  
     syphilis of, ii, 174; iii, 632  
     tuberculosis of, i, 363; iii, 631  
     in typhoid fever, i, 86  
 Pancreatic apoplexy, iii, 610  
     juice, ferments of, iii, 289  
         secretion of, iii, 288  
 Pancreatitis, chronic, iii, 629  
     experimental, iii, 612  
     in mumps, i, 921  
     suppurative, iii, 622  
     traumatic, iii, 612  
     in typhoid fever, i, 120  
 Papillary cystadenoma of kidney, iii, 1002  
 Papillitis, v, 326  
     lingualis, iii, 62  
 Papilloma of brain, v, 315  
 Papular eruption in syphilis, ii, 164  
 Paracentesis abdominis, iii, 667  
     in cirrhosis of liver, iii, 524, 534  
     of pericardium, iv, 56  
     thoracic, cardiac insufficiency and, iv, 196  
 Paracentral convolutions, tumors of, v, 332  
 Parachordodes, ii, 296  
 Paradoxical embolism, iv, 363, 414, 553  
 Paraffin embolism, iv, 562  
 Paragonimiasis, ii, 226  
 Paragonimus westermanii, ii, 226  
 Paragordius, ii, 296  
 Paragraphia, v, 66, 75  
 Paragusia, iii, 43  
 Paralactic acid, ii, 599  
 Paralysis in abscess of brain, v, 381  
     in acute infective polyneuritis, v, 590  
     agitans, v, 764  
         ductless glands and, v, 767  
         exophthalmic goitre and, iv, 870  
     Paralysis agitans, parathyroid glands and, iv, 903; v, 767  
         trauma and, v, 764  
     in amaurotic family idiocy, v, 869  
     in apoplexy, v, 438, 440  
     in arsenic poisoning, ii, 376  
     ascending bulbar, v, 798  
     asthenic, v, 798  
     atrophic, in syringomyelia, v, 221  
     in beriberi, ii, 495  
     birth, brachial, v, 552  
     of brachial plexus, v, 550  
     in brain tumor, v, 337  
     of cardiac nerves, v, 522  
     in cerebrospinal syphilis, v, 825  
     of cervical plexus, v, 543  
     of circumflex nerve, v, 546  
     compression, in syphilis, ii, 162  
     diaphragmatic, v, 544  
     in diphtheria, i, 701  
     diver's, ii, 345  
     Duchenne-Erb, v, 551  
     Erb's syphilitic, v, 246  
     of external rectus muscle, v, 485  
     of facial nerve, v, 495  
         in hysteria, v, 679  
     flaccid, v, 178  
     of gastric nerves, v, 523  
     glosso-labio-laryngeal, v, 113  
     of gluteal nerves, v, 558  
     in hematomyelia, v, 260  
     in hysteria, v, 650, 678  
     in infantile convulsions, v, 795  
     of inferior oblique muscle, v, 485  
         rectus muscle, v, 485  
     of internal rectus muscle, v, 485  
     of intestines in acute peritonitis, ii, 681  
         obstruction from, iii, 418  
     in intradural hemorrhage, v, 258  
     of intra-ocular muscles, v, 486  
     Klumpke's, v, 552  
     Landry's, v, 247  
     laryngeal, ii, 867; v, 518, 521  
     in lead poisoning, ii, 365  
     in lesion of cerebellum, v, 286  
         of crura cerebri, v, 281  
         of internal capsule, v, 276  
         of mid-brain, v, 283  
         of pons, v, 289  
         of tegmentum, v, 281  
     of long thoracic nerve, v, 544  
     of lower arm, v, 552  
     of lumbar plexus, v, 560  
     of median nerve, v, 548  
     in mercury poisoning, ii, 379  
     Millard-Gubler type, v, 290  
     in multiple neuritis, v, 580  
     of muscles, v, 68  
     of musculocutaneous nerve, v, 546  
     of musculospiral nerve, v, 547  
     of nerves of legs, v, 556  
     of ocular nerves, v, 483  
     of oesophageal nerves, v, 523  
     of oesophagus, iii, 90  
     periodic, v, 809  
     in pernicious anemia, iv, 631  
     of pharyngeal nerves, v, 517

- Paralysis of phrenic nerve, v, 543  
 in polycythemia, iv, 685  
 of popliteal nerves, v, 559  
 in progressive spinal muscular atrophy, v, 84  
 pseudo-bulbar, v, 290, 978  
 of pulmonary nerves, v, 523  
 of radial nerve, v, 547  
 of sacral plexus, v, 560  
 of sciatic nerve, v, 558, 560  
 in smallpox, i, 813  
 of soft palate, iii, 62  
 "soul," v, 268  
 spastic, v, 179  
   spinal, v, 101  
     hereditary, v, 105  
 of spinal accessory nerve, v, 524  
 of superior oblique muscle, v, 486  
   rectus muscle, v, 485  
 of suprascapular nerve, v, 545  
 in syphilitic myelitis, v, 240  
 of tongue, iii, 62; v, 530  
 in traumatic neuroses, v, 651  
 of trigeminal nerve, v, 549  
 in tumor of thyroid gland, iv, 833  
 unilateral, v, 108  
 of upper arm, v, 551  
 in uremia, iii, 836  
 of vagus nerve, v, 517
- Paralytic conjugate deviation in apoplexy, v, 439  
 phenomena in chorea, v, 780  
   in migraine, v, 716  
 rabies, i, 965
- Paralyzing vertigo, v, 515
- Paramimia, v, 66
- Paramnesia, v, 65
- Paramyoclonus multiplex, v, 807
- Paranephric abscess, iii, 970
- Paranephritis, iii, 970  
 in tuberculosis of kidney, iii, 980
- Paranoia, trauma and, v, 647
- Paraphasia, v, 66
- Paraplegia in abscess of brain, v, 381  
 in aortic embolism, iv, 566  
   thrombosis, iv, 566  
 in arteriosclerosis, iv, 461  
 in cerebrospinal syphilis, v, 825  
 in combined sclerosis, v, 127  
 in diabetes mellitus, ii, 707  
 Erb's syphilitic, v, 246  
 in hysteria, v, 679  
 spastic, in Hodgkin's disease, iv, 767  
 in syphilitic myelitis, v, 241  
 in tumors of spinal cord, v, 203
- Parapleural echinococcus, ii, 1066
- Parapraxia, v, 66
- Parasites, animal, ii, 277  
 appendicitis and, iii, 360  
 embolism and, iv, 558  
 inflammation of thoracic duct and, iv, 584  
 intestinal, in lobar pneumonia, i, 254  
   pernicious anemia and, iv, 623  
 in kala-azar, iv, 965  
 kidney infections and, iii, 952  
 of lymph nodes, iv, 753
- Parasites, malarial, ii, 56, 59, 78  
 of myocardium, iv, 142  
 of thoracic duct, iv, 589
- Parasitic aneurism, iv, 472  
 cysts of brain, v, 318  
   of peritoneum, iii, 733  
   of spleen, iv, 953
- Parasyphilis, ii, 185  
 of nervous system, v, 822
- Parathyroid glands, iv, 898  
 eclampsia and, iv, 903  
 epilepsy and, iv, 903  
 in exophthalmic goitre, iv, 857  
 kidneys and, iv, 904  
 myasthenia and, iv, 903  
 myoclonus and, iv, 903  
 myotonia and, iv, 903  
 osteomalacia and, iv, 903; v, 949  
 paralysis agitans and, iv, 903  
 pellagra and, iv, 903  
 rickets and, iv, 903  
 tetany and, iv, 901; v, 750, 753  
 thyroid gland and, iv, 903  
 tumors of, iv, 904  
 insufficiency, iv, 901
- Paratyphoid fever, i, 198
- Paravertebral triangle of dulness in sero-fibrinous pleuritis, ii, 1023
- Parenchymatous changes in kidney, iii, 774  
 degeneration of liver, iii, 466  
 goitre, iv, 839  
 hepatitis, iii, 494  
 multiple neuritis, v, 577  
 myocarditis, iv, 113
- Paresis in diabetes mellitus, ii, 707  
 general, v, 850  
   tabes dorsalis, and, v, 843  
   trauma and, v, 647  
 in lead poisoning, ii, 368  
 migraine and, v, 720  
 neuralgia and, v, 727
- Paretic gait, v, 73
- Parietal lobe, tumors of, v, 333
- Parkinson's disease, v, 764
- Parorexia, iii, 134
- Parotid gland in mumps, i, 920
- Parotitis, acute, iii, 69  
 in bacillary dysentery, i, 776  
 chronic, iii, 69  
 epidemic, i, 919  
 gastric ulcer and, iii, 189  
 in glandular fever, i, 1013  
 in lobar pneumonia, i, 263  
 in rheumatic fever, i, 999  
 in subphrenic pyopneumothorax, iii, 712  
 in typhoid fever, i, 103
- Paroxysmal tachycardia, iv, 300
- Pasteur immunization in rabies, i, 970
- Patellar tendon reflex, v, 77
- Patency of ductus arteriosus Botalli, iv, 409
- Patent foramen ovale, iv, 355
- Pectoral heart, iv, 346
- Pediculoides ventricosus, ii, 322
- Pediculosis, ii, 326
- Pediculus, ii, 326

- Peliomata in typhoid fever, i, 101  
 Peliolosis rheumatica, iv, 701  
 Pellagra, ii, 472; v, 139  
     parathyroid glands and, iv, 903  
 Pelvic peritonitis, kidney infections and, iii, 952  
 Pelvis in endemic cretinism, iv, 882  
     of kidney, tumors of, iii, 1006  
 Pemphigoid rash in typhoid fever, i, 101  
 Pemphigus in congenital syphilis, ii, 189  
     of mouth, iii, 54  
     pellagrosus, ii, 483  
     in vaccination, i, 843  
 Pentastomum constriction, ii, 324  
     denticulatum cyst of spleen, iv, 954  
 Pentosuria, ii, 600, 712; iii, 822  
 Peptic ulcer, iii, 168  
     jejunal, iii, 349  
     of cesophagus, iii, 84  
 Peptonuria, iii, 820  
 Perception, anomalies of, v, 37  
     sense, v, 18  
 Periaortitis, acute, iv, 452  
 Periarthritis nodosa, iv, 470  
     nodular, ii, 181  
 Peri-appendicular abscess, iii, 364, 370, 377  
 Peri-articular arthritis deformans, v, 908  
 Pericardial effusion, iv, 49  
     sac, obliteration of, hypertrophy of heart and, iv, 181  
 Pericarditic pseudocirrhosis of liver, iii, 488, 716; iv, 60, 63, 64  
 Pericarditis, iv, 41  
     in abscess of lung, ii, 995  
     acute, iv, 43  
     adhesive, iv, 44, 58  
     in arthritis deformans, v, 914  
     in cerebrospinal meningitis, i, 609  
     chorea and, iv, 42  
     chronic, iv, 51  
     perihepatitis and, iii, 492  
     circumscribed, iv, 42  
     with effusion, iv, 49  
     in endocarditis, iv, 161  
     gonorrhoeal, i, 752; iv, 42  
     in gout, ii, 749  
     in lobar pneumonia, i, 257  
     in measles, i, 907  
     mediastinitis and, iv, 59, 62  
     purulent, iv, 50  
     in rheumatic fever, i, 988; iv, 41  
     in scarlet fever, i, 877  
     serofibrinous, iv, 50  
     sicca, iv, 50  
     tricuspid insufficiency and, iv, 247  
     tuberculous, iv, 42, 66  
     in typhoid fever, i, 126  
     in uremia, iii, 839  
 Pericardium, absence of, iv, 77, 344  
     adherent, iv, 58  
     cardiac insufficiency and, iv, 199  
     anomalies of, iv, 344  
     carcinoma of, iv, 74  
     defects of, iv, 77, 344  
     diverticulum of, iv, 346  
     effusions into, cardiac insufficiency and, iv, 193  
 Pericardium, fibroids of; iv, 76  
     hernia of, iv, 346  
     hydatids of, iv, 76  
     lymphoma of, iv, 76  
     neoplasms of, iv, 74  
     paracentesis of, iv, 56  
     sarcoma of, iv, 74  
     syphilis of, iv, 76  
     tuberculosis of, i, 359; iv, 66  
     unattached, iv, 345  
 Perichondritis, gonococcal, i, 760  
     in lobar pneumonia, i, 248  
 Pericolitis, iii, 697  
 Pericolitis, iii, 354  
     acute, iii, 694  
 Perigastric abscess, iii, 696  
     adhesions in stenosis of pylorus, iii, 262  
 Perigastritis in gastric ulcer, iii, 815  
 Perihepatitis, iii, 486, 488, 718  
 Perimesosigmoiditis, iii, 715  
 Perinephric abscess, iii, 970  
 Perinephritis, iii, 970  
     in tuberculosis of kidney, iii, 980  
 Periodic paralysis, v, 809  
 Periodontitis, iii, 56  
 Periosteal dyscrasia, v, 978  
 Periostitis alveolaris dentalis, iii, 56  
     in syphilis, ii, 163  
 Peripheral arteries, embolism of, iv, 568  
     thrombosis of, iv, 568  
     nerves, diseases of, v, 533  
     neuritis, v, 573  
     venous thrombosis, iv, 539  
 Peripleuritis, ii, 1051  
 Perisigmoiditis, iii, 695  
 Perisplenic peritonitis, iv, 943  
 Perisplenitis, iv, 943  
     in abscess of spleen, iv, 945  
 Peristalsis in cirrhosis of stomach, iii, 277  
     in intestinal colic, iii, 304  
     of intestines, iii, 286  
     in stenosis of pylorus, iii, 271  
 Peristaltic unrest, iii, 151  
 Peritoneal abscesses, iii, 698  
 Peritoneum, absorption from, iii, 655  
     angioma of, iii, 733  
     in cirrhosis of liver, iii, 518  
     cysts of, iii, 733  
     fibroma of, iii, 730  
     fibromyoma of, iii, 730  
     gangliated neuroma of, iii, 731  
     immunity of, iii, 657  
     lipoma of, iii, 732  
     myoma of, iii, 730  
     myxoma of, iii, 733  
     pseudomyxoma of, iii, 740  
     retroperitoneal hematoma of, iii, 738  
     tuberculosis of, i, 364; iii, 720  
     tumors of, iii, 730, 740  
 Peritonitis, acute circumscribed, iii, 693  
     diffuse, iii, 670  
     in amoebic dysentery, ii, 32, 44  
     in appendicitis, iii, 364, 372, 374  
     in bacillary dysentery, i, 776  
     chronic diffuse, iii, 715  
     simple, iii, 713

- Peritonitis deformans, iii, 718  
     fibrosa, iii, 718  
     from intussusception, iii, 410  
     gonococcal, i, 763; iii, 686  
     in lobar pneumonia, i, 254  
     lymphadenitis and, iv, 736  
     pelvic, kidney infections and, iii, 952  
     perisplenic, iv, 943  
     pneumococcal, iii, 685  
     progressive fibrinopurulent, iii, 676  
     puerperal, iii, 686  
     in rheumatic fever, i, 997  
     in scarlet fever, i, 878  
     in suppurative pancreatitis, iii, 625  
     tuberculous, i, 364; iii, 720  
     in typhoid fever, i, 116  
 Peritonsillar abscess, ii, 849  
 Perityphlitis, iii, 355  
 Peri-ureteritis, iii, 974  
     kidney infections and, iii, 952  
     in pyelitis, iii, 954  
 Pernicious anemia, iv, 619  
     malarial fevers, ii, 80  
 Persistent truncus arteriosus, iv, 375  
 Petechiæ in smallpox, i, 799  
     in toxic gastritis, iii, 234  
     in tuberculosis, i, 419  
 Petit mal, v, 604  
 Pfeiffer's reactions in Asiatic cholera, i, 679  
 Pflüger's theory of protein metabolism, ii, 592  
 Phagocytosis, i, 57  
 Phantom tumor, iii, 299  
 Pharyngeal diphtheria, i, 696  
     nerves, paralysis of, v, 517  
     tonsil, hypertrophy of, ii, 840  
     tuberculosis, i, 429  
 Pharyngitis, ii, 843, 844, 845, 846  
 Pharynx, diseases of, ii, 842  
     in myasthenia gravis, v, 804  
     cedema of, ii, 847  
     in scarlet fever, i, 863  
     in typhoid fever, i, 104  
 Phimosis, kidney infections and, iii, 952  
 Phlebitis, iv, 574  
     acute suppurative, iv, 577  
     chronic, iv, 575  
     in glands, i, 651  
     gonorrhœal, i, 752  
     in gout, ii, 749; iv, 574  
     idiopathic, iv, 574  
     migrans, iv, 531, 576  
     phlebosclerosis and, iv, 576  
     in rheumatic fever, i, 990  
     in scarlet fever, i, 878  
     septic, iv, 574  
     suppurative, of hepatic veins, iii, 460  
     syphilitic, iv, 576  
     traumatic, iv, 574  
     tuberculous, iv, 576  
     in typhoid fever, i, 128, 132  
 Phlebosclerosis, phlebitis and, iv, 576  
 Phlegmonous cholecystitis, iii, 563  
     oesophagitis, iii, 80  
     pharyngitis, ii, 846  
 Phloridzin diabetes, ii, 685  
 Phobias, v, 52  
 Phonic spasm, v, 522  
 Phosphaturia, iii, 814  
 Phosphorus poisoning, ii, 381  
 Photopsia, v, 36  
 Phrenic nerve in aneurism of aorta, iv, 501  
     paralysis of, v, 543  
 Phthisis, acute, i, 424  
     florida, i, 344, 354  
 Phthirus pubis, ii, 326  
 Physaloptera, ii, 293  
 Phytin, metabolism of, ii, 562  
 Phytotoxins, i, 34  
 Pick's disease, iii, 488, 716  
 Pigmentation in Addison's disease, iv, 789,  
     790, 792, 795  
     in arsenic poisoning, ii, 375  
     in exophthalmic goitre, iv, 866  
     in Hodgkin's disease, iv, 767  
     of lymph nodes, iv, 746  
     in scleroderma, iv, 1019  
     in silver poisoning, ii, 385  
     in splenic anemia, iv, 958  
     of thymus, iv, 912  
     in typhoid fever, i, 102  
 Pineal body, iv, 819  
     in hypopituitarism, iv, 817  
     lesion of, v, 291  
     tumor of, v, 336  
 Pinworm infection, ii, 291  
 Pithiatism, v, 659  
 Pituitary body, iv, 833  
     in acromegaly, iv, 808  
     carcinoma of, iv, 804  
     circulatory changes in, iv, 804  
     degeneration of, iv, 804  
     diabetes and, ii, 683, 722  
     lesion of, v, 291  
     sarcoma of, iv, 804  
     tumors of, iv, 804  
         hypertrophy of heart and, iv,  
         113  
     dystrophy, iv, 815  
 Plague, i, 620  
     bubonic, i, 632  
     fulminating, i, 634  
     intestinal, i, 635  
     pestis minor, i, 632  
     septicemic, i, 634  
 Plasmodium falciparum, ii, 56, 59, 92, 102  
     quotidianum, ii, 57, 92, 102  
     malariae, ii, 56, 58, 92, 102  
     vivax, ii, 56, 57, 92, 102  
 Plastic bronchitis, ii, 907  
     pleuritis, ii, 1005  
 Pleura, actinomycosis of, ii, 1050  
     carcinoma of, ii, 1061  
     diseases of, ii, 1004  
     echinococcus disease of, ii, 1065  
     lipoma of, ii, 1061  
     nocardiosis of, ii, 1051  
     sarcoma of, ii, 1064  
     syphilis of, ii, 1051  
     tuberculosis of, i, 356  
         pericarditis and, iv, 42  
     tumors of, ii, 1060, 1061, 1064  
 Pleural echinococcus, ii, 1065  
     fluids in carcinoma, ii, 1063

- Pleural fluids, chyliform, ii, 1057  
 chylous, ii, 1057  
 pseudochylous, ii, 1057
- Pleuritic adhesions, hypertrophy of heart and, iv, 181
- Pleuritis, ii, 1004  
 in abscess of lung, ii, 995  
 acute fibrinous, ii, 1005  
   purulent, ii, 1040  
   serofibrinous, ii, 1010  
 in aneurism of, aorta, iv, 508  
 in arthritis deformans, v, 915  
 bronchiectasis and, ii, 914  
 in bronchopneumonia, ii, 964  
 chronic, ii, 1051  
   perihepatitis and, iii, 492  
   peritonitis and, iii, 716  
   with effusion, ii, 1052  
 in cirrhosis of liver, iii, 526  
 defects of pericardium and, iv, 345  
 diaphragmatic, ii, 1049  
 encysted, ii, 1049  
 fibrinous, ii, 1005  
 gonococcal, i, 762  
 hemorrhagic, ii, 1053  
 infectious (non-tuberculous) ii, 1011  
 interlobar, ii, 1050  
 in lobar pneumonia, i, 248  
 in malarial fevers, ii, 86  
 metapneumonic, ii, 1012  
 pericarditis and, iv, 42  
 plastic, ii, 1005  
 in rheumatic fever, i, 996  
 in scarlet fever, i, 876  
 in subphrenic pyopneumothorax, iii, 712  
 in tuberculosis, i, 430; ii, 1010  
 tumors of lung and, ii, 1001  
 in typhoid fever, i, 123  
 in typhus fever, i, 930  
 in whooping cough, i, 660  
 with effusion, ii, 1010
- Pleurodynia, v, 945
- Pneurogenous interstitial pneumonia, ii, 970
- Plexiform neuromata, v, 571
- Plumbism, ii, 355
- Pneumatosi, iii, 150
- Pneumatia, iii, 806
- Pneumococcus, i, 208, 274  
 in empyema, ii, 1041
- Pneumogastric nerve, diseases of, v, 517
- Pneumokoniosis, ii, 976  
 cirrhosis of liver and, iii, 510
- Pneumonia, abortive, i, 237  
 acute endocarditis and, iv, 153  
   meningitis and, v, 168  
 apical, i, 236  
 aspiration, i, 206; ii, 958  
 asthenic, i, 236  
 bronchiectasis and, ii, 914, 917  
 caseous, i, 352, 424  
 catarrhal, ii, 957  
 central, i, 237  
 in cerebrospinal meningitis, i, 599, 609  
 in childhood, i, 238  
 deglutition, ii, 958
- Pneumonia in diabetes mellitus, ii, 706  
 in erysipelas, i, 587  
 gangrene of lung and, ii, 989  
 gelatinous, i, 352  
 in glanders, i, 652  
 gonococcal, i, 762  
 inhalation, i, 206  
 lobar, i, 202  
   defect of pericardium and, iv, 345  
   in diphtheria, i, 266  
   in malarial fevers, i, 266  
   in measles, i, 265  
   in pregnancy, i, 264  
   in scarlet fever, i, 265  
   in typhoid fever, i, 122, 207, 267  
   in typhus fever, i, 267  
 lobular, ii, 957  
 in malarial fevers, ii, 86  
 pericarditis and, iv, 42  
 in plague, i, 628, 634  
 pleurogenous interstitial, ii, 970  
 postoperative, i, 237  
 in rheumatic fever, i, 996  
 in scarlet fever, i, 878  
 senile, i, 236  
 terminal, i, 237  
 in tuberculosis, i, 350, 424  
 tumors of lung and, ii, 1001  
 white, ii, 172
- Pneumonic fever, i, 202
- Pneumonitis, i, 202
- Pneumopericardium, iv, 71
- Pneumothorax, ii, 1071  
 abscess of lung and, ii, 1074  
 acutissimus, ii, 1084  
 artificial, ii, 1084  
 bronchiectasis and, ii, 1074  
 closed, ii, 1084  
 defect of pericardium and, iv, 345  
 double, ii, 1084  
 gangrene of lung and, ii, 1074  
 infarction of lung and, ii, 1074  
 in influenza, i, 542  
 in lobar pneumonia, i, 250  
 open, ii, 1083  
 partial, ii, 1084  
 thoracentesis and, ii, 1038, 1074  
 in tuberculosis, i, 430, 512; ii, 1073  
 valvular, ii, 1083  
 wounds of chest and, ii, 1074
- Pneumotiphoid, i, 90
- Poikilocytosis in pernicious anemia, iv, 637
- Poisoning, alcohol, ii, 396  
 arsenic, ii, 373  
 brass, ii, 386  
 carbon monoxide, ii, 388  
 cardiac insufficiency and, iv, 199, 202  
 cheese, ii, 467  
 cocaine, ii, 445  
 copper, ii, 386  
 by fish, ii, 454  
 food, ii, 450  
 illuminating gas, ii, 388  
 lead, ii, 355, 369  
   arteriosclerosis and, iv, 455  
 manganese, ii, 386  
 meat, ii, 458, 461, 462



- Poisoning, mercury, ii, 378  
 methyl alcohol, ii, 398  
 milk, ii, 466  
 nitric oxide, ii, 395  
 opium, ii, 433  
 phosphorus, ii, 381  
 shell-fish, ii, 455  
 silver, ii, 384  
 tetany and, v, 749  
 tin, ii, 386  
 vegetable, ii, 468  
 zinc, ii, 386
- Poliencephalitis, acute inferior, v, 368  
 hemorrhagic, ii, 429  
 superior, v, 365
- Poliencephalomyelitis, v, 369
- Poliomyelitis, acute, i, 733  
 chronic anterior, v, 82
- Pollantin, ii, 862, 865
- Pollen antitoxin, ii, 861  
 toxin, ii, 859
- Polonium, rays from, ii, 339
- Polyarteritis acuta nodosa, iv, 470
- Polyarthritis, multiple neuritis and, v, 585
- Polychromates, v, 25
- Polycythemia, iv, 684  
 of congenital cyanosis, iv, 337, 340  
 in patent ductus arteriosus, iv, 415  
 tuberculosis of spleen and, iv, 684
- Polydipsia in diabetes mellitus, ii, 698  
 in tumor of brain, v, 338
- Polyglandular syndrome, iv, 818
- Polygraphy, heart-beat and, iv, 80
- Polyideic somnambulism, v, 44
- Polymyositis hemorrhagica, v, 879
- Polyneuritic psychosis, v, 45
- Polyneuritis, v, 573  
 acute infective, v, 590  
 in arsenic poisoning, ii, 376  
 facial paralysis and, v, 497  
 gonococcal, i, 761  
 recurring, v, 591
- Polyneuclear cells in lymphoid leukemia, iv, 673
- Polyorrhmenitis, iii, 488; iv, 60, 63, 64
- Polyplasmia in chlorosis, iv, 653
- Polyps of appendix, iii, 386
- Polysymptomatic neuroses, iii, 164
- Polyuria, 791, 806  
 in acromegaly, iv, 808  
 in brain tumor, v, 337, 338, 342  
 in chronic interstitial nephritis, iii, 900  
 pyelitis, iii, 961  
 in diabetes, ii, 698, 725  
 in exophthalmic goitre, iv, 868  
 in hypopituitarism, iv, 816  
 in typhoid fever, i, 141
- Pons, lesion of, v, 289  
 tumors of, v, 336
- Pontile syndrome, v, 289
- Pontomedullocerebellar space, lesion of, v, 289
- Popliteal nerves, paralysis of, v, 559
- Porencephaly, apoplexy and, v, 409  
 epilepsy and, v, 598
- Pork-measly tapeworm, ii, 249
- Porocephaliasis, ii, 323
- Portal cirrhosis of liver, iii, 508  
 pyemia, i, 566; iii, 463  
 vein, abscess of liver and, iii, 497  
 embolism of, iii, 463  
 perforation of, in gastric ulcer, iii, 186  
 in splenic anemia, iv, 955  
 thrombosis of, iii, 460; iv, 544
- Position, anomalies of, v, 30
- Posterior basic meningitis, i, 615  
 spinal sclerosis, v, 833
- Posthemoplegic chorea, v, 466
- Posthemorrhagic anemia, acute, iv, 655
- Postnasal catarrh, ii, 838
- Postoperative pneumonia, i, 237  
 thrombosis, iv, 528
- Pott's disease, v, 188
- Pox, ii, 144
- Precipitin test in glanders, i, 654
- Precipitins, i, 57
- Pregnancy, acute yellow atrophy of liver and, iii, 472  
 apoplexy and, v, 423, 425  
 arthritis deformans and, v, 902  
 asthma and, ii, 933  
 cholelithiasis and, iii, 571  
 chorea and, v, 777, 779  
 diabetes mellitus and, ii, 677  
 exophthalmic goitre and, iv, 851  
 kidney of, iii, 867  
 chronic nephritis and, iii, 893  
 infections and, iii, 952  
 myasthenia gravis and, v, 799  
 necrosis of pancreas and, iii, 612  
 nephritis and, iii, 856  
 osteomalacia and, v, 948  
 pernicious anemia and, iv, 621  
 progressive spinal muscular atrophy and, v, 81  
 pyelonephritis of, iii, 961  
 serodiagnosis of, ii, 652  
 syphilis and, ii, 187  
 toxemia of, myocarditis and, iv, 108  
 tuberculosis of kidney and, iii, 978  
 of peritoneum and, iii, 721
- Presbyopia, v, 36
- Pressure, sense of, v, 19, 32  
 ulcers of oesophagus, iii, 84
- Priapism in myeloid leukemia, iv, 665
- Primitive aortic arches, development of, iv, 329
- Proctitis, gonorrhœal, i, 747
- Professional neuroses, v, 741
- Profeta's immunity in syphilis, ii, 153
- Progeria, iv, 817
- Progressive bulbar palsy, v, 113  
 muscular dystrophy, v, 117  
 spinal muscular atrophy, v, 82, 87
- Propulsion, v, 74
- Propyl alcohol, ii, 400
- Prostate, carcinoma of, iii, 1038  
 diseases of, iii, 1026  
 hypertrophy of, iii, 1035  
 kidney infections and, iii, 952  
 tuberculosis of, i, 366
- Prostatitis, chronic, i, 746; iii, 1032

- Protein equilibrium, ii, 560  
 fat formation and, ii, 604  
 foreign, ii, 651  
 metabolism, ii, 579  
 microbic, i, 33  
 sparing, ii, 583
- Protopathic sensibility, v, 17
- Protozoa, diarrhoea and, iii, 325  
 pathogenic, i, 19
- Protozoön in scarlet fever, i, 860, 863
- Pruritus in catarrhal cholangitis, iii, 553  
 in diabetes mellitus, ii, 704  
 in exophthalmic goitre, iv, 866  
 in Hodgkin's disease, iv, 768  
 in jaundice, iii, 452
- Psammoma of brain, v, 318
- Psammoma sarcoma of brain, v, 315
- Pseudamphistomum truncatum, ii, 233
- Pseudo-achondroplasia, v, 969
- Pseudo-angina, iv, 293
- Pseudobulbar paralysis, v, 290, 798
- Pseudochylous fluids, ii, 1058
- Pseudocirrhosis, pericardial, iii, 488, 716;  
 iv, 60
- Pseudocrisis in lobar pneumonia, i, 240
- Pseudohallucinations, v, 37
- Pseudohypertrophic progressive muscular  
 atrophy, v, 122
- Pseudoleukemia, iv, 681, 772
- Pseudomembranous bronchitis, ii, 907  
 pyelitis, iii, 954
- Pseudomyxoma of peritoneum, iii, 740
- Pseudoparalytica, v, 798
- Pseudopellagra, ii, 485
- Pseudoperforation of gastric ulcer, iii, 188
- Pseudosclerosis, v, 158
- Pseudotabes, syphilitic, v, 826
- Pseudotrichinae, ii, 302
- Pseudotubercle bacilli, i, 297
- Psilosis, iii, 393; ii, 45
- Psittacosis, i, 1024
- Psoriasis syphilide, ii, 189
- Psoriasis linguæ, iii, 64  
 of mouth, iii, 53  
 syphilitica, ii, 165  
 in vaccination, i, 843
- Psychasthenia, v, 816
- Psychic blindness, v, 299  
 epilepsy, v, 602  
 vomiting, iii, 144
- Psychical neuralgia, v, 726
- Psychoneuroses, digestion and, iii, 37
- Psychoses in hysteria, v, 689  
 traumatic, v, 642  
 in tuberculosis, i, 321, 392  
 in typhoid fever, i, 133
- Psychosis, Korsakow's, ii, 406, 427  
 polyneuritic, v, 45  
 post-epileptic, v, 603
- Ptomain poisoning, ii, 510  
 nephritis and, iii, 853
- Ptomains, i, 553
- Ptosis in cerebrospinal syphilis, v, 825  
 in hemiplegia, v, 459  
 in hysteria, v, 679  
 in ocular paralysis, v, 488  
 in paralyzing vertigo, v, 515
- Ptosis in tuberculosis, i, 423
- Ptyalism, iii, 68
- Pudendal plexus, neuralgia of, v, 740
- Puerperal eclampsia, acute yellow atrophy  
 of liver and, iii, 473  
 infections, chronic peritonitis and, iii,  
 714  
 peritonitis, iii, 686
- Pulex irritans, ii, 325
- Pulmonary anthrax, i, 646  
 artery, accessory, iv, 439  
 anomalies of, iv, 439  
 dilatation of, iv, 398  
 defects of interauricular sep-  
 tum and, iv, 359  
 embolism of, iv, 562  
 left subclavian artery from, iv,  
 436  
 aspergillosis, i, 1058  
 atresia, congenital, iv, 388  
 diphtheria, i, 706  
 distomatosis, ii, 226  
 endarteritis, acute, iv, 413  
 hypertrophic osteo-arthropathy, i, 411;  
 v, 981  
 insufficiency, iv, 255  
 nerves, paralysis of, v, 523  
 œdema in rheumatic fever, i, 996  
 roundworms, ii, 302  
 stasis, iv, 19  
 stenosis, iv, 258  
 congenital, iv, 388  
 in tuberculosis, i, 433  
 thrombosis, iv, 547  
 cardiac insufficiency and, iv, 197  
 tuberculosis, acute, i, 424  
 miliary, i, 419  
 chronic, i, 426  
 latent, i, 427  
 subacute, i, 426  
 veins, anomalies of, iv, 441
- Pulsus alternans, iv, 187  
 paradoxus in adherent pericardium,  
 iv, 62
- Pupillary reaction, hemianopic, v, 482  
 of Wernicke, v, 271  
 reflexes, anomalies of, v, 76
- Pupils in apoplexy, v, 439  
 in disseminated sclerosis, v, 152  
 in general paresis, v, 856  
 in tabes dorsalis, v, 837
- Purins, ii, 733  
 in protein metabolism, ii, 589
- Purpura, iv, 687  
 abdominalis, iv, 708  
 angioneurotic œdema and, iv, 707,  
 999, 1002  
 in cerebrospinal meningitis, iv, 688  
 chronic, iv, 713  
 continuous, iv, 713  
 erythema and, iv, 705  
 fulminans, iv, 712  
 hemorrhagica, iv, 711  
 aplastic anemia and, iv, 699  
 Henoch's, iv, 701  
 idiopathic, iv, 691  
 intermittent, iv, 713

Purpura, iodine and, iv, 689  
 mechanical, iv, 690  
 nervous, iv, 690  
 orthostatique, iv, 690  
 in Raynaud's disease, iv, 990  
 rheumatic fever and, iv, 704  
 rheumatica, iv, 701  
 secondary, iv, 688  
 senile, iv, 688  
 simplex, iv, 700  
 in smallpox, iv, 688  
 toxic, iv, 689  
 in tuberculosis, iv, 688  
 in typhus fever, iv, 688  
 Purpuric smallpox, i, 804  
 Pustular eruption in syphilis, ii, 166  
 Putrid bronchitis, ii, 911  
 Pyelitis, iii, 937  
   acute catarrhal, iii, 953  
   in children, iii, 961  
   chronic, iii, 954  
   croupous, iii, 954  
   cystic, iii, 954  
   diphtheritic, iii, 954  
   fibrinous, iii, 954  
   gangrenous, iii, 954  
   granular, iii, 954  
   pseudomembranous, iii, 954  
   in typhoid fever, i, 143  
 Pyelonephritis, iii, 937  
   of pregnancy, iii, 961  
 Pyelonephrosis, iii, 937  
 Pyemia, i, 551  
   in bronchiectasis, ii, 917  
   endocarditis and, iv, 161  
   inflammation of thoracic duct and, iv, 582  
   myocarditis and, iv, 109  
   nephritis and, iii, 850  
   portal, i, 566; iii, 463  
 Pylephlebitis, suppurative, i, 570; iii, 463  
 Pylethrombosis, iv, 544  
 Pylorospasm, iii, 149, 262  
   acute dilatation and, iii, 25  
   congenital, iii, 269  
 Pylorus, incontinence of, iii, 159  
   stenosis of, iii, 262  
     gastric ulcer and, iii, 189  
 Pyogenic infections of kidney, iii, 937  
 Pyonephrosis, iii, 957  
   renal calculus and, iii, 1018  
 Pyopneumopericardium, iv, 71  
 Pyopneumothorax, subphrenic, iii, 710  
 Pyorrhœa alveolaris, iii, 56  
   arthritis deformans, and, v, 901  
   in tuberculosis, i, 431  
 Pyroplasma hominis, i, 1021  
 Pyrosis in acute gastritis, iii, 228  
 Pyuria, iii, 805  
   in typhoid fever, i, 142

## Q

QUADRIGEMINAL bodies, lesion of, v, 282  
 tumor of, v, 282

Quartan malarial parasite, ii, 56, 58  
 Quinke's disease, iv, 998  
 Quinsy, ii, 849

## R

RABIES, i, 956  
 Radial nerve, paralysis of, v, 547  
 Radium, goitre and, iv, 838  
   rays from, ii, 339  
 Rales in tuberculosis, i, 408, 441  
 Ranula, iii, 52  
   pancreatica, iii, 640  
 Rat-bite disease, i, 1030  
 Rat trypanosome, ii, 116  
 Raynaud's disease, iv, 975  
   arthritis deformans and, v, 929  
 Reaction, anomalies of, v, 48  
   of degeneration in amyotrophic lateral sclerosis, v, 99  
   distal, v, 535  
   myasthenic, v, 73  
   myotonic, v, 73, 891  
 Rectum, injuries of, kidney infections and, iii, 952  
   syphilis of, ii, 175  
   in typhoid fever, i, 117  
   ulcers of, in tuberculosis, i, 432  
 Rectus muscles, paralysis of, v, 485  
 Recurrent endocarditis, iv, 148  
   laryngeal nerve in aneurism of aorta, iv, 502  
 Reflex dyspnoea, iv, 38  
   neuralgia, v, 726  
   vomiting, iii, 145  
 Regurgitation, gastric, iii, 140  
 Relapsing fevers, ii, 133  
 Remittent fever, ii, 133  
 Renal arteries, anomalies of, iii, 786  
   embolism of, iv, 564  
   calculus, iii, 1007  
   casts, iii, 798  
   colic, renal calculus and, iii, 1019  
   pelvis, empyema of, iii, 937  
   system in cardiac insufficiency, iv, 199, 210  
     in typhoid fever, i, 140  
     vessels, thrombosis of, iii, 789; iv, 542  
 Residual peritoneal abscesses, iii, 698  
 Resistance, sensation of, v, 17  
 Respiratory centre, ii, 812  
   accessory, ii, 815  
   influenza, i, 539  
   irregularity, heart beat and, iv, 83  
   movements in pericarditis, iv, 48  
   quotient, ii, 618  
   stridor in bronchial lymphadenitis, iv, 735  
     in tuberculosis of bronchial glands, iv, 743  
   system in acute endocarditis, iv, 153  
   in angioneurotic edema, iv, 1007  
   in arthritis deformans, v, 915  
   in cardiac insufficiency, iv, 206  
   in chlorosis, iv, 651  
   in exophthalmic goitre, iv, 868  
   in gout, ii, 745

- Respiratory system in lead poisoning, ii, 364  
 in pernicious anemia, iv, 634  
 in rheumatic fever, i, 995  
 syphilis of, ii, 170  
 tuberculosis of, i, 347  
 in typhoid fever, i, 80, 120  
 tract, obstruction of, ii, 815, 818
- Retina in chronic nephritis, iii, 910  
 lesions of, v, 474
- Retinal arteries, embolism of, iv, 570  
 thrombosis of, iv, 570  
 hemorrhages in chronic interstitial nephritis, iii, 911  
 in leukemia, iv, 665, 671  
 in pernicious anemia, iv, 632
- Retinitis, v, 474  
 albuminuric, iii, 911; v, 475  
 syphilitic, v, 473
- Retrobulbar neuritis, v, 37, 479
- Retroperitoneal abscesses, iii, 698  
 subphrenic abscess and, iii, 701  
 cysts of peritoneum, iii, 738  
 hematoma, iii, 738  
 sarcoma, iii, 742
- Retropharyngeal abscess, ii, 846; iv, 734
- Retropulsion, v, 74
- Revaccination, i, 837
- Rhabditis, ii, 295  
 cornwalli, ii, 302  
 genitalis, ii, 316  
 mellyi, ii, 303  
 terrisola, ii, 302
- Rhabdomyoma, congenital, iv, 351
- Rhachitis annularis, v, 978
- Rhagades in congenital syphilis, ii, 189
- Rheumatic fever, i, 974  
 acute endocarditis and, iv, 154  
 aneurism and, iv, 475  
 angioneurotic edema and, iv, 999  
 aortic insufficiency in, iv, 221  
 aortitis in, iv, 450, 475  
 cerebral, i, 992  
 chorea and, v, 775  
 dilatation of heart in, iv, 49  
 endocarditis and, iv, 156  
 exophthalmic goitre and, iv, 851  
 lymphadenitis in, iv, 737  
 myocarditis and, iv, 108, 118  
 nephritis and, iii, 850  
 pericarditis and, iv, 41  
 purpura and, iv, 704  
 thrombosis in, iv, 529  
 tricuspid insufficiency and, iv, 244
- Rheumatoid arthritis, v, 897, 908
- Rhinitis in influenza, i, 539
- Rhinogenic abscess of brain, v, 385
- Rhythmic segmentation of intestines, iii, 286
- Ribs in achondroplasia, v, 957  
 cervical, v, 554
- Rickets, ii, 775  
 achondroplasia and, v, 966  
 annular, v, 955  
 congenital, ii, 795; v, 955  
 convulsions and, v, 792, 793  
 fetal, v, 955
- Rickets, infantile scurvy and, ii, 807  
 intra-uterine, v, 955  
 late, ii, 796  
 micromelic, v, 955  
 parathyroid glands and, iv, 903  
 syphilis and, ii, 777  
 thymus and, iv, 924  
 tuberculosis and, i, 322
- Rickety rosary, ii, 783
- Riga's disease, iii, 66
- Rigg's disease, iii, 56
- Rinné's test in nerve deafness, v, 510
- Rocky Mountain spotted fever, i, 1020
- Romberg symptom in tabes dorsalis, v, 838
- Rose fever, ii, 859  
 spots in typhoid fever, i, 100
- Roseola in syphilis, ii, 164
- Rotch's sign in congenital cyanosis, iv, 337
- Round ulcer, iii, 168
- Roundworm infection, ii, 270
- Roundworms, intestinal, ii, 271, 297  
 muscular, ii, 297  
 pulmonary, ii, 302  
 subcutaneous, ii, 302
- Rubella, i, 912
- Rumination, iii, 138
- Rupture of aorta, iv, 487  
 of cerebral arteries, v, 400  
 of heart, hemopericardium and, iv, 70  
 insufficiency and, iv, 193  
 of cesophagus, iii, 85  
 of spleen, iv, 947  
 of thoracic duct, iv, 586  
 of valves of heart, aortic insufficiency and, 219, 227

## S

- SACCULED aneurism, iv, 479, 492
- Sacral disseminated sclerosis, v, 154  
 plexus, paralysis of, v, 560
- Sacro-iliac joint in arthritis deformans, v, 912, 923
- St. Anthony's fire, i, 585
- St. Gothard tunnel disease, ii, 271
- St. Vitus' dance, v, 774
- Saliva, iii, 67
- Salivary calculi, iii, 71  
 gland, atrophy of, iii, 74  
 enlargement of, iii, 73  
 stone in, iii, 71  
 swelling of, iii, 70  
 syphilis of, ii, 174; iii, 71  
 tuberculosis of, i, 363; iii, 70  
 tumors of, iii, 72  
 secretion, iii, 68
- Salivation, iii, 48, 68
- Salpingitis, thoracic duct and, iv, 583
- Salvarsan in cerebrospinal syphilis, v, 830  
 intraspinal, v, 831, 862  
 mercury and, ii, 214  
 in relapsing fever, ii, 142  
 in syphilis, ii, 211
- Salvarsanized serum, v, 831, 862
- Sand, intestinal, iii, 295
- Sanguinous cysts of peritoneum, iii, 738

- Sarcinæ in carcinoma of stomach, iii, 213  
 Sarcoma of adrenal glands, iv, 784  
   of appendix, iii, 387  
   of brain, v, 313, 314  
   of intestine, iii, 431  
   of kidney, iii, 1003  
   of liver, iii, 546  
   of lung, ii, 998  
   of lymph nodes, iv, 747  
   of lymphatic vessels, iv, 601  
   of mediastinum, ii, 1093  
   of œsophagus, iii, 105  
   of pancreas, iii, 650  
   of pericardium, iv, 74  
   of pituitary body, iv, 804  
   of pleura, ii, 1064  
   retroperitoneal, iii, 742  
   of spinal cord, v, 197  
   of spleen, iv, 951  
   of thoracic duct, iv, 587  
   of thymus, iv, 932  
   of thyroid gland, iv, 832  
 Sarcoptes, ii, 318, 319  
 Sarcoptic ascariasis, ii, 318  
   itch, ii, 318  
 Saturnism, ii, 355  
 Scanning speech, v, 74  
 Scapulodynia, v, 945  
 Scarlet fever, i, 856  
   acute endocarditis in, iv, 156  
   in animals, i, 860  
   apoplexy and, v, 417  
   arteriosclerosis and, iv, 454  
   bronchopneumonia and, ii, 958  
   diphtheria and, i, 700  
   multiple neuritis and, v, 575  
   myocarditis and, iv, 108  
   nephritis and, iii, 849  
   purpura fulminans and, iv, 712  
   surgical, i, 884  
   tuberculosis and, i, 321  
   typhoid fever and, i, 158  
   vaccination and, i, 843, 887  
   whooping cough and, i, 659  
 Schistosoma, ii, 239, 240  
 Schizonts, ii, 63  
 Schönlein's disease, iv, 701  
 Schwabach's test, v, 24  
 Sciatic nerve, paralysis of, v, 558, 560  
   in spondylitis deformans, v, 927  
 Sciatica, v, 562  
   gonococcal, i, 761  
 Scirrhus of thyroid gland, iv, 833  
 Scleroderma, diffuse, iv, 1014  
   Raynaud's disease and, iv, 986,  
   1018, 1021  
   of mouth, iii, 54, 60  
 Sclerosis, amyotrophic lateral, v, 89  
   of brain, v, 140  
   of cerebellum, v, 287  
   of coronary arteries, myocarditis and,  
   iv, 109  
   diffuse, v, 160  
   disseminated, v, 140, 154  
   insular, v, 140  
   lateral, v, 101  
   of mediastinal glands, ii, 1111  
 Sclerosis, miliary, v, 161  
   multiple, v, 140  
   posterior spinal, v, 833  
   subacute combined, of cord, v, 126  
   tuberous, v, 160  
 Scoleciasis, ii, 328  
 Scotoma, v, 34  
 Scotomata in migraine, v, 712, 714  
 Scorbutus, ii, 799  
 Screw-worm, ii, 328  
 Scrofula, i, 357  
   of lymph nodes, iv, 745  
 Scrofuloderma, i, 374  
 Scrofulosis, tuberculosis and, i, 320  
 Scurvy, ii, 799  
   infantile, ii, 804  
   rickets and, ii, 807  
 Secretion, iii, 19, 30, 290  
   of saliva, iii, 67  
 Secretory functions, anomalies of, v, 79  
 Segmentation of myocardium, iv, 116  
 Semilunar cusps, anomalies of, iv, 400  
   defective development of, iv, 403  
 Seminal vesicles, tuberculosis of, i, 366  
 Semple's immunization in rabies, i, 977  
 Senile aortic insufficiency, iv, 219  
   dementia, trauma and, v, 647  
   emphysema of lungs, ii, 982  
   heart, myocarditis and, iv, 126  
   pneumonia, i, 236  
   purpura, iv, 689  
 Sensation, deep, v, 536  
   disturbances of, v, 28  
   epicritic, v, 537  
   protopathic, v, 537  
 Sense of acceleration, v, 18  
   of equilibrium, v, 11  
   of fatigue, v, 19, 32  
   of hearing, v, 22  
   of hunger, v, 19, 31  
   interior, v, 17  
   of movement, v, 30  
   muscle, v, 17  
   pain, v, 17  
   perception, v, 18, 30  
   of position, v, 30  
   of pressure, v, 19, 32  
   of resistance, v, 17, 30  
   sexual, v, 19, 32  
   of sight, v, 24  
   of smell, v, 21, 33  
   of taste, v, 21, 33  
   temperature, v, 20, 32  
   of thirst, v, 19, 32  
   of touch, v, 19, 32  
   vestibular, v, 18, 30  
   vibration, v, 18, 30  
 Sensibility, deep, v, 17, 184  
   anomalies of, v, 30  
   epicritic, v, 17, 184  
   mesoblastic, v, 17  
   anomalies of, v, 30  
   osseous, v, 30  
   protopathic, v, 17, 184  
   stereognostic, v, 18, 30  
 Sensory aphasia, v, 74, 292, 296  
   area of cortex of brain, v, 265

- Sensory disturbances in occupation neuroses, v, 747  
 phenomena in tetany, v, 759
- Sepsis, gastro-intestinal, pernicious anemia and, iv, 623  
 oral, iii, 41
- Septic anemia, acute, iv, 658  
 endocarditis, chronic, iv, 148  
 phlebitis, iv, 574
- Septicemia, i, 551  
 acute endocarditis and, i, 558  
 in anthrax, i, 557  
 in appendicitis, iii, 372  
 in bubonic plague, i, 558  
 in diphtheria, i, 555  
 in gonorrhœa, i, 556, 749  
 inflammation of thoracic duct and, iv, 582  
 in influenza, i, 544, 555  
 in pneumonia, i, 555  
 in pulmonary insufficiency, iv, 257  
 in rheumatic fever, i, 558  
 in scarlet fever, i, 864  
 septic anemia and, iv, 658  
 in typhoid fever, i, 557
- Septicemic plague, i, 634
- Septicopyemia, endocarditis and, iv, 157
- Septicopyemic abscess of liver, iii, 499
- Septum, aortic, defects of, iv, 374, 377  
 development of, iv, 327  
 of heart, absence of, complete, iv, 370  
 anomalies of, iv, 352  
 development of, iv, 370  
 interauricular, defects of, iv, 355  
 interventricular, defects of, iv, 363  
 development of, iv, 327
- Sera, nephritis and, iii, 852
- Serofibrinous exudate in pericarditis, iv, 43  
 pericarditis, iv, 50  
 pleuritis, ii, 1000
- Serositis, multiple, iii, 488; iv, 60, 63, 64
- Serous cysts of spleen, iv, 951  
 meningitis, ii, 415
- "Serum disease," i, 62, 729
- Setaria equina, ii, 315
- Shaking palsy, v, 764
- Shell-fish poisoning, ii, 455
- Sialodochitis fibrinosa, iii, 70
- Sialorrhœa, iii, 35, 68
- Sialosis, iii, 68
- Siberian liver-fluke disease, ii, 233
- Sick headache, v, 709
- Siderosis, ii, 976
- Sight, anomalies of, v, 34  
 sense of, v, 24
- Silicosis, hepatic, iii, 470
- Silver poisoning, ii, 384
- Simulation in hysteria, v, 659, 667
- Sino-auricular node, iv, 79
- Sinus irregularities, iv, 83  
 thrombosis, abscess of brain and, v, 378  
 of Valsalva, aneurism of, iv, 492
- Siriasis, ii, 333
- Sitophobia, iii, 158, 165
- Skin in acromegaly, iv, 807
- Skin in acute catarrhal cholangitis, iii, 553  
 nephritis, iii, 864  
 in Addison's disease, iv, 789, 791, 792  
 in angioneurotic œdema, iv, 1005  
 in arsenic poisoning, ii, 375  
 in arthritis deformans, v, 916  
 atrophy of, from x-rays, ii, 336  
 in cardiac insufficiency, iv, 209  
 in chronic interstitial nephritis, iii, 906  
 in diabetes mellitus, ii, 704  
 in diffuse scleroderma, iv, 1016, 1018  
 in diseases of peripheral nerves, v, 540  
 in endemic cretinism, iv, 882, 884  
 eruptions, gonococcal, i, 764  
 in erythromelalgia, iv, 1028  
 in exophthalmic goitre, iv, 866  
 exudative lesions of, iv, 1003  
 in facial hemiatrophy, v, 992  
 in herpes zoster, v, 572  
 in Hodgkin's disease, iv, 767  
 in hypopituitarism, iv, 815  
 in hysteria, v, 686  
 infections, endocarditis and, iv, 153, 157  
 in influenza, i, 545  
 in jaundice, iii, 451  
 in kala-azar, iv, 970, 971  
 in lead poisoning, ii, 361  
 in leprosy, i, 527  
 in lobar pneumonia, i, 259  
 in lymphoid leukemia, iv, 672, 676  
 in measles, i, 907  
 in meningitis, i, 599, 603, 609  
 in myxœdema, iv, 890, 891  
 in neuralgia, v, 728, 734  
 in oidiomycosis, i, 1052  
 in opium smoking, ii, 436  
 in pellagra, ii, 478  
 in plague, i, 623  
 in polymyositis hemorrhagica, v, 879  
 in Raynaud's disease, iv, 990  
 reflexes in meningitis, i, 607  
 in rheumatic fever, i, 996  
 in scarlet fever, i, 863, 872  
 in silver poisoning, ii, 385  
 in splenic anemia, iv, 958  
 in sunstroke, ii, 331  
 in syringomyelia, v, 224  
 tuberculosis of, i, 373, 386  
 in typhoid fever, i, 99
- Skull in achondroplasia, v, 956  
 in cerebrospinal meningitis, i, 610, 613  
 in endemic cretinism, iv, 882  
 hyperostosis of, v, 988  
 in Paget's disease, v, 971  
 percussion of, in acute miliary tuberculosis, i, 423
- Sleeping sickness, ii, 119, 120, 128
- Smallpox, i, 783  
 acute endocarditis and, iv, 156  
 arteriosclerosis and, iv, 454  
 confluent, i, 803  
 discrete, i, 803  
 experimental, i, 793  
 fetal, i, 810  
 hemorrhagic pustular, i, 804

- Smallpox inoculation, i, 792  
 lymphadenitis in, iv, 737  
 measles and, i, 907  
 nephritis and, iii, 849  
 purpuric, i, 804; iv, 688
- Smell, anomalies of, v, 33  
 centre of, v, 271  
 disturbances of, v, 471  
 in hysteria, v, 684  
 sense of, v, 21, 33
- Smoker's patch, iii, 64
- Sneezing, mechanism of, ii, 827
- Snuffles in congenital syphilis, ii, 188
- Softening of adrenal glands, iv, 783
- Somatic teniasis, ii, 263
- Somatopsyche, v, 42
- Somnambulisms, v, 44
- "Soul paralysis," v, 268
- Sparganum, ii, 263, 270
- Spargosis, iv, 596
- Spasm, habit, v, 784  
 of muscles of neck, v, 525  
 of tongue, v, 532  
 phonic, v, 522  
 of tongue, iii, 62
- Spasmodic torticollis, v, 526
- Spasmophilia, v, 604
- Spastic constipation, iii, 313  
 dysphonia, v, 522  
 gait in pernicious anemia, iv, 630  
 paraplegia in Hodgkin's disease, iv, 767  
 spinal paralysis, v, 101, 105  
 syringomyelia, v, 220  
 tabes, v, 101
- Spastic-paretic gait, v, 73
- Speech, anomalies of, v, 74
- Sphincters in multiple neuritis, v, 584
- Spina bifida, hydrocephalus and, v, 354
- Spinal accessory nerve, paralysis of, v, 524  
 ataxic gait, v, 73  
 caries, v, 188  
 cord, abscess of, v, 254  
 in Addison's disease, iv, 789  
 anemia of, v, 255  
 in aneurism of aorta, iv, 505  
 carcinoma of, v, 196  
 in cerebrospinal meningitis, i, 597  
 combined system diseases of, v, 125  
 compression of, v, 188  
 diseases of, v, 178, 187  
 fissure of, v, 210  
 hemorrhage into, v, 260  
 hyperemia of, v, 257  
 lesions of, v, 178, 180  
 myeloma of, v, 198  
 in pellagra, ii, 478  
 in pernicious anemia, iv, 625  
 in Raynaud's disease, iv, 989  
 in rheumatic fever, i, 994  
 sarcoma of, v, 191  
 sclerosis of, combined, v, 139  
 segmental localization of, v, 178  
 syphilis of, v, 823  
 thrombosis of, v, 256  
 tumors of, v, 196
- Spinal cord, vascular disorders of, v, 255  
 curvature in syringomyelia, v, 222  
 disseminated sclerosis, v, 154  
 membranes, hemorrhage into, v, 257  
 meningitis, localized serous, v, 174  
 muscular atrophy, progressive, v, 82, 87  
 nerves, diseases of, v, 542  
 paralysis, spastic, v, 101, 105  
 syphilitic, v, 828  
 roots, diseases of, v, 567  
 sclerosis, posterior, v, 833
- Spine, deformities of, acute dilatation of  
 stomach and, iii, 251  
 in Paget's disease, v, 971  
 tuberculosis of, subphrenic abscess  
 and, iii, 701
- Spirillum fever, ii, 133
- Spirochæta duttoni, ii, 136  
 pallida, ii, 144  
 in general paresis, v, 851, 852  
 in syphilis of nervous system, v, 823  
 in tabes dorsalis, v, 835  
 recurrentis, ii, 133  
 refringens, ii, 146, 148
- Spirochetes in mouth, iii, 41
- Spirometry in tuberculosis, i, 404
- Splanchnoptosis, iii, 747  
 in chlorosis, iv, 651  
 digestion and, iii, 35  
 gastro-intestinal, iii, 754  
 hepatic, iii, 754  
 lithemic, iii, 754  
 pregnancy and, iii, 751
- Spleen, abscess of, iv, 944  
 subphrenic abscess and, iii, 700  
 absence of, iv, 938  
 accessory, iv, 938  
 in acute hepatitis, iii, 495  
 infective polyneuritis, v, 590  
 yellow atrophy of liver, iii, 476  
 in Addison's disease, iv, 794  
 in alcoholism, ii, 404  
 amyloid, iv, 949  
 kidney and, iii, 935  
 liver and, iii, 469  
 angioma cavernosum of, iv, 950  
 in aplastic anemia, iv, 645  
 in arthritis deformans, v, 909, 915  
 carcinoma of, iv, 951  
 of liver and, iii, 544  
 in catarrhal cholangitis, iii, 554  
 in cerebrospinal meningitis, i, 599  
 in chlorosis, iv, 651  
 in chronic peritonitis, iii, 718  
 in cirrhosis of liver, iii, 517, 523, 538  
 in congenital syphilis, ii, 190  
 congestion of, iv, 940  
 cysts of, iv, 951  
 descent of, iii, 758  
 in diphtheria, i, 694  
 dislocated, iv, 938  
 in enlargement of thymus, iv, 916  
 enzymes and, iv, 935  
 in exophthalmic goitre, iv, 857, 867  
 fibroma of, iv, 950

- Spleen, floating, iv, 938  
 in Gaucher's splenomegaly, iv, 962  
 in glandular fever, i, 1013  
 gumma of, ii, 176  
 in Henoch's purpura, iv, 710  
 in hepatic thrombosis, iv, 546  
 in Hodgkin's disease, iv, 762, 767  
 infarction of, iv, 564, 946  
 infectious diseases and, iv, 936  
 inflammation of, iv, 940  
 intoxications and, iv, 936  
 in kala-azar, iv, 970, 971  
 large, iv, 938  
 in lobar pneumonia, i, 233  
 lobulated, iv, 938  
 lymphangioma of, iv, 950  
 in lymphoid leukemia, iv, 672  
 in malarial fevers, ii, 71, 78  
 malformations of, iv, 937  
 malpositions of, iv, 938  
 metabolism of, iv, 935  
 movable, iv, 938  
 movements of, iv, 934  
 multiple, iv, 938  
 in myeloid leukemia, iv, 662, 665  
 nerve supply of, iv, 934  
 in pernicious anemia, iv, 626, 634  
 in plague, i, 628  
 in polycythemia, iv, 685  
 portal blood and, iv, 936  
 in purpura hemorrhagica, iv, 711  
 in rickets, ii, 790  
 rupture of, iv, 947  
 sarcoma of, iv, 951  
 in scarlet fever, i, 865  
 in septicemia, i, 562  
 small, iv, 938  
 in smallpox, i, 790  
 in splenic anemia, iv, 955  
 syphilis of, ii, 176  
 tuberculosis of, i, 369  
   polycythemia and, iv, 684  
 tumors of, iv, 942, 950  
 in typhoid fever, i, 85, 117  
 in typhus fever, i, 926  
 vessels of, thrombosis of, iv, 546  
 wandering, iv, 938
- Splenectomy in pernicious anemia, iv, 644
- Splenic anemia, iv, 954  
 artery, embolism of, iv, 554  
 fever, i, 639  
 vein, thrombosis of, iv, 948  
   in necrosis of pancreas, iii, 619
- Splenitis, acute suppurative, iv, 944  
 capsular, iv, 943
- Splenomegaly, Gaucher's, iv, 962  
 in splenic anemia, iv, 957  
 tropical febrile, iv, 964
- Splenoptosis, iv, 938
- Splits of intima of aorta, iv, 487
- Spondylitis deformans, v, 923  
 in typhoid fever, i, 147
- Sporadic cretinism, iv, 883
- Sporoblasts, ii, 63
- Sporont, ii, 62
- Sporotrichosis, i, 1055
- Sporotrichosis, disseminated gummatous  
   i, 1056  
   localized, i, 1057
- Sporozoites, ii, 63
- Sprue, ii, 45; iii, 393
- Stammering, v, 74
- Staphylococcus in empyema, ii, 1041  
 epidermidis albus in peritonitis, iii, 674  
 pyogenes albus in peritonitis, iii, 674  
   aureus in encephalitis, v, 363  
   in peritonitis, iii, 675  
   in mouth, iii, 40
- Starvation, metabolism of, ii, 606
- Stasis in irritable heart, iv, 319  
 vascular, iv, 20  
 venous, in pericarditis, iv, 21, 48
- Status epilepticus, v, 609  
 hemicranus in migraine, v, 718  
 lymphaticus, iv, 912, 916  
   thymic death and, iv, 921  
 thymicolymphaticus, iv, 916
- Steapsin, iii, 289
- Steatorrhœa in calculus of pancreas, iii, 635  
 in cyst of pancreas, iii, 645  
 in diseases of pancreas, iii, 599
- Stenosing tuberculosis of intestines, iii, 390
- Stenosis, aortic, iv, 228  
 congenital, iv, 391, 399  
 of duodenum, iii, 262  
 of intestines, congenital, obstruction  
   from, iii, 408  
 left-sided conus, iv, 400  
 mitral, iv, 239  
   congenital, iv, 406  
   defects of interauricular septum  
     and, iv, 381  
 of œsophagus, iii, 96  
 pulmonary, iv, 258  
   congenital, iv, 388  
 of pylorus, iii, 262  
   congenital hypertrophic, iii, 269  
 subaortic, iv, 398  
 tricuspid, iv, 250  
   congenital, iv, 403
- Stercoral ulcers, iii, 351
- Stereognosis, v, 18, 30
- Sternoclavicular joint in arthritis deformans, v, 911
- Still's disease, v, 917, 931
- Stokes-Adams syndrome, iv, 90, 209, 305  
 in aortic stenosis, iv, 231  
 arteriosclerosis and, iv, 306  
 bulbar origin of, iv, 309  
 bundle of His and, iv, 308  
 heart-block and, iv, 306  
 in myocarditis, iv, 127  
 syphilis and, iv, 308  
 vagus and, iv, 307
- Stomach in Addison's disease, iv, 789  
 in alcoholism, ii, 404  
 anacidity of, iii, 153  
 atony of, iii, 160  
   cholelithiasis and, iii, 161  
 carcinoma of, iii, 199  
   colloid, iii, 203  
   multiple, iii, 202  
   pernicious anemia and, iv, 627



- Stomach, carcinoma of, scirrhus, iii, 203  
 secondary, iii, 201  
 simplex, iii, 203  
 squamous-celled, iii, 203  
 cardia of, carcinoma of, iii, 220  
 spasm of, iii, 147  
 cirrhosis of, iii, 241, 273  
 circumscribed, iii, 276  
 dilatation of, acute, iii, 24, 248  
 chronic, iii, 253  
 gastropnoxis and, iii, 254  
 idiopathic, iii, 253  
 in lobar pneumonia, i, 254  
 nephropnoxis and, iii, 756  
 obstructive, iii, 255  
 symptomatic, iii, 255  
 in tuberculosis, i, 502  
 diseases of, functional, iii, 106  
 organic, iii, 168  
 distension of, iii, 150  
 cardiac arrhythmia and, iv, 287  
 disturbance of function of, iii, 23  
 hemorrhagic erosions of, iii, 838  
 hyperacidity of, iii, 112  
 hyperchlorhydria of, iii, 112  
 hypochlorhydria of, iii, 152  
 lesser curvature of, carcinoma of, iii, 219  
 motor insufficiency of, iii, 247  
 neuroses of, iii, 106  
 perforation of, iii, 619  
 pylorus of, incontinence of, iii, 159  
 spasm of, iii, 149  
 stenosis of, iii, 262  
 subacidity of, iii, 152  
 syphilis of, ii, 174  
 tuberculosis of, i, 361, 431  
 in typhoid fever, i, 104  
 ulcer of, iii, 168  
 arteriosclerosis and, iv, 464  
 Stomatitis, aphthous, iii, 46  
 catarrhal, iii, 45  
 in chronic intestinal nephritis, iii, 904  
 gangrenous, iii, 49  
 gonorrhoeal, i, 748; iii, 51  
 herpetic, iii, 51  
 hypomycetic, iii, 50  
 in measles, i, 906  
 mercurial, iii, 48  
 mycotic, iii, 50  
 in tuberculosis, i, 431  
 ulcerative, iii, 47  
 in uremia, iii, 838  
 Strabismus in ophthalmoplegia, v, 484  
 in tuberculosis, i, 423  
 Strangulation of intestines, iii, 394  
 hemorrhage and, iii, 307  
 Streptococcus brevis in mouth, iii, 40  
 in empyema, ii, 1041  
 erysipelatis, i, 585  
 pyogenes in acute peritonitis, iii, 674  
 in appendicitis, iii, 361  
 in mouth, iii, 40  
 rheumaticus, i, 975  
 in scarlet fever, i, 859, 864  
 Streptothrichosis, i, 1045  
 Stricture of intestines, iii, 394  
 of intestines, obstruction from, iii, 407  
 of cesophagus, non-malignant, iii, 96  
 Strongyloides stercoralis, ii, 285, 286  
 Strongyloidosis, ii, 285  
 Strongylus gibsoni, ii, 294  
 Struma, iv, 835  
 Stupor, v, 47, 63, 65  
 Stuttering, v, 74  
 Subacidity, iii, 152  
 Subacute combined sclerosis of spinal cord, v, 126  
 Subaortic stenosis, iv, 398  
 Subclavian artery, embolism of, iv, 567  
 left, from pulmonary artery, iv, 436  
 right, from descending thoracic aorta, iv, 436  
 thrombosis of, iv, 567  
 Submaxillary glands in typhoid fever, i, 104  
 Suboxidation, ii, 515, 543, 546  
 Subphrenic abscess, iii, 699  
 left anterior intraperitoneal, iii, 703  
 extraperitoneal, iii, 705  
 posterior intraperitoneal, iii, 705  
 in lobar pneumonia, i, 253  
 right anterior, iii, 703  
 extrahepatic, iii, 705  
 without gaseous contents, iii, 699  
 pneumothorax, ii, 1089  
 pyopneumothorax, iii, 710  
 appendicular abscess and, iii, 711  
 duodenal ulcer and, iii, 710  
 gastric ulcer and, iii, 710  
 hydatid cyst of liver and, iii, 711  
 Succus entericus, iii, 290  
 Succussion sound in pneumothorax, ii, 1080  
 Suffocative catarrh, ii, 962  
 Suffocation, mechanics of, ii, 835  
 Sugar in urine, iii, 821  
 in diabetes mellitus, ii, 701  
 Sugar-content of blood, ii, 597  
 Sugar-iced liver, iii, 488  
 Sulphur, metabolism of, ii, 561  
 Sunstroke, ii, 331  
 Sun-traumatism, ii, 333  
 Superior vena cava, thrombosis of, iv, 541  
 Superoxidation, ii, 518, 543, 546  
 Suppurative cholangitis, iii, 556  
 nephritis, iii, 937  
 pancreatitis, iii, 622  
 phlebitis of hepatic veins, iii, 460  
 pyelonephritis, iii, 937  
 pyelophlebitis, i, 570; iii, 463  
 Suprarenal gland in chronic interstitial nephritis, iii, 899  
 diseases of, iv, 777  
 Suprascapular nerve, paralysis of, v, 545  
 Surgical kidney, iii, 937  
 Surra, ii, 117  
 Sweating sickness, i, 1017  
 Sydenham's chorea, v, 774

- Sympathetic nerves in aneurism of aorta,  
     iv, 501  
   system, iii, 24, 35, 107  
     Addison's disease and, iv, 788  
     in exophthalmic goitre, iv, 857
- Symptomatic anemia, iv, 654
- Syndrome, Benedikt's, v, 369
- Syndrome, cerebellar deficit, v, 287
- Millard-Gubler, v, 369
- pontile, v, 289  
     superior, v, 291  
   tegmental, v, 281  
   thalamic, v, 279, 334, 467  
   Weber's, v, 369
- Synechia cordis, iv, 23
- Synesthesia, v, 29
- Synovial membrane in arthritis deformans,  
   v, 904, 905
- Syphilides, cutaneous, ii, 150  
     pigmented, ii, 186  
     tuberculous, ii, 166  
     ulcerated, ii, 166
- Syphilis, ii, 144  
     achondroplasia and, v, 967  
     acute yellow atrophy of liver and, iii,  
       473  
     of adrenal glands, iv, 786  
     of alimentary canal, ii, 174  
     amyloid degeneration and, ii, 152  
       kidney and, iii, 933  
       spleen and, iv, 949  
     amyotrophic lateral sclerosis and, v, 90  
     aneurism and, iv, 474  
     aortic insufficiency and, iv, 219, 222  
     apoplexy and, v, 423, 425, 426, 427  
     of arteries, ii, 181  
     arteriosclerosis and, iv, 454; v, 393  
     of brain, v, 823  
     of bronchi, ii, 170, 906  
     cerebral, aneurism and, v, 399  
       diabetes insipidus and, ii, 722  
       thrombosis and, v, 400, 401  
     cerebrospinal, v, 823, 824  
     chancre in, ii, 154  
     chorea and, v, 775, 777  
     chronic perihepatitis and, iii, 489  
     of circulatory organs, ii, 180  
     cirrhosis of liver and, iii, 509  
     congenital, ii, 187  
     dilatation of aorta and, iv, 485  
     epilepsy and, v, 595  
     exophthalmic goitre and, iv, 851  
     fetal, ii, 187  
     general paresis and, v, 850  
     gumma in, ii, 150  
     of heart, ii, 183  
     hereditary hemiplegia in, v, 418  
     hypertrophic osteo-arthritis and,  
       v, 982  
     inflammation of spleen and, iv, 941  
       of thoracic duct and, iv, 584  
     insomnium, ii, 156  
     of intestine, ii, 175; iii, 391  
     of kidneys, ii, 179  
     of larynx, ii, 871  
     leukoplakia and, iii, 64  
     of liver, ii, 176; iii, 541
- Syphilis of lung, ii, 171  
     of lymph nodes, iv, 745  
     of lymphatic vessels, iv, 596  
     of mouth, iii, 58  
     myelitis and, v, 228  
     myocarditis and, iv, 112  
     of myocardium, iv, 142  
     nephritis and, iii, 851  
     of nervous system, v, 822  
     neurasthenia and, v, 629  
     in ocular paralysis, v, 488  
     of oesophagus, ii, 174; iii, 86  
     osteitis deformans and, v, 973  
     of pancreas, ii, 174; iii, 632  
     of pericardium, iv, 76  
     pernicious anemia and, iv, 622  
     of pleura, ii, 1051  
     postnatal infantile hemiplegia and, v,  
       418  
     pregnancy and, ii, 187  
     of rectum, ii, 175  
     of respiratory system, ii, 170  
     of salivary glands, ii, 174; iii, 71  
     secondary, ii, 157, 168  
     of spinal cord, v, 823  
     of spleen, ii, 176  
     Stokes-Adams syndrome and, iv, 308  
     of stomach, ii, 174  
     syringomyelia and, v, 212  
     tabes dorsalis and, v, 833  
     tertiary, ii, 169, 184  
     of thoracic duct, iv, 585  
     thrombosis in, iv, 530  
     of thymus, iv, 929  
     of thyroid gland, iv, 831  
     of trachea, ii, 170, 906  
     tuberculosis and, i, 320, 436  
     typhoid fever and, i, 159  
     vaccination and, i, 844
- Syphilitic aortitis, ii, 182; iv, 474  
     arteritis, ii, 181  
     choroidoretinitis, v, 475  
     endocarditis, ii, 183  
     myelitis, v, 237, 828  
     myocarditis, ii, 184  
     paraplegia, Erb's, v, 246  
     phlebitis, iv, 576  
     retinitis, v, 475  
     spinal paralysis, v, 828  
     vitiligo, ii, 186
- Syphiloderma, gummatous, ii, 184  
     tuberculous, ii, 184  
     ulcerative, ii, 185
- Syphiloma, ii, 149  
     of brain, v, 320, 321
- Syringomyelia, v, 211
- Systemic veins, anomalies of, iv, 439
- Systolic retraction in adherent pericar-  
   dium, iv, 61

## T

- TABES amaurotica, v, 844  
     atactica, v, 844  
     dolorosa, v, 844  
     dorsalis, v, 833

- Tabes dorsalis, exophthalmic goitre and,  
     iv, 870  
     migraine and, v, 720  
     neuralgia and, v, 727  
     syphilis and, v, 833  
     incomplete, v, 844  
     juvenilis, v, 844  
     mesenterica, i, 359; iv, 744  
     paralytica, v, 844  
     paresthetica, v, 844  
     spastic, v, 101  
     superior, v, 840, 844  
     visceralis, v, 844  
 Tâche cérébrale in meningitis, i, 601  
 Tachycardia, iv, 90, 279  
     in defects of interauricular septum, iv,  
         362  
     in exophthalmic goitre, iv, 860  
     gastric distension and, iv, 312  
     irregular, iv, 93  
     orthostatic, iv, 304  
     paroxysmal, iv, 85, 281, 300  
         bradycardia and, iv, 302  
     in patent ductus arteriosus, iv, 415  
     primary, iv, 280  
     reflex, iv, 280, 312, 315  
     regular, iv, 93  
     sexual organs and, iv, 318  
     simple, iv, 87  
     suspension of inhibition and, iv, 314  
     in syphilis of myocardium, iv, 143  
     in tabes, iv, 277  
     in tuberculosis, i, 500  
     vagus and, iv, 276, 280, 304  
 Tænia, ii, 246, 249, 252, 264, 265  
 Tæniasis, ii, 258  
 Talalgia, i, 760  
 Tapeworm, broad, ii, 255  
     dwarf, ii, 252  
     fat, ii, 247  
     infection, ii, 245  
     pork, ii, 249  
 Tartar, iii, 71  
 Taste, disturbances of, iii, 43  
     sense of, v, 21  
 Tauraman, i, 305  
 Tay-Sachs disease, v, 863  
 Teeth, carious, in tuberculosis, i, 431  
     in endemic cretinism, v, 882  
     Hutchinson's, iii, 59  
     in lead poisoning, ii, 360  
     in myxœdema, iv, 891  
     in rickets, ii, 782  
 Tegmental syndrome, v, 281  
 Tegmentum, lesion of, v, 281  
     tumors of, v, 336  
 Telangiectasis in cirroid aneurism, iv, 487  
 Temporal lobe, tumors of, v, 331  
 Tendons, tuberculosis of, i, 373  
 Tenesmus in amœbic dysentery, ii, 37  
     in diarrhœa, iii, 326  
     in intussusception, iii, 411  
 Teniasis, ii, 245, 263  
 Tenosynovitis, gonocœcal, i, 760  
 Teratoma of mediastinum, ii, 1101  
     of thymus, iv, 933  
 Terminal infections, i, 559  
 Terminal pneumonia, i, 237  
 Ternidens deminutus, ii, 294  
 Tertian malarial parasite, ii, 56, 57  
 Testicle in smallpox, i, 791  
     tuberculosis of, i, 366  
 Testicular neuralgia, v, 740  
 Tetanolysin, i, 34  
 Tetanospasmin, i, 34  
 Tetanus, i, 575, 579, 580  
 Tetany, v, 748  
     auto-intoxication and, ii, 511  
     benign, v, 759  
     in dilatation of stomach, iii, 257  
     epidemic, v, 750, 752  
     gastric ulcer and, iii, 189  
     hysteria and, v, 677  
     idiopathic, v, 749  
     in lobar pneumonia, i, 262  
     non-operative, iv, 902  
     operative, iv, 901  
     osteomalacia and, v, 949  
     parathyreopriva, v, 761  
     parathyroid glands and, iv, 901; v,  
         749, 750, 753  
     in pylorospasm, iii, 150  
     in rickets, ii, 788  
     in typhoid fever, i, 136, 158  
     vaccination and, i, 844  
 Tetranychus molestissimus, ii, 323  
 Thalamic syndrome, v, 279, 334, 467  
 Thermanesthesias, v, 32  
 Thirst, sense of, v, 19, 32  
 Thomsen's disease, v, 886  
 Thoracentesis, ii, 1035  
     emphysema and, ii, 1039  
     pneumothorax and, ii, 1038, 1074  
 Thoracic abscess in tuberculosis, i, 435  
     actinomycosis, i, 1039  
     duct, absence of, iv, 580  
         anomalies of, iv, 580  
         carcinoma of, iv, 588  
         chyle cysts of, iv, 586  
         ectasia of, iv, 585  
         endothelioma of, iv, 588  
         fat embolism and, iv, 582  
         hemorrhage into, iv, 581  
         inflammation of, iv, 582  
         lymphangivitis of, iv, 584  
         obstruction of, iv, 586  
         parasites of, iv, 589  
         rupture of, iv, 586  
         sarcoma of, iv, 587  
         syphilis of, iv, 585  
         thrombosis of, iv, 581  
         tuberculosis of, iv, 584  
         tumors of, iv, 587  
     nerve, long, paralysis of, v, 544  
     wall, perforation of, in purulent pleu-  
         ritus, ii, 1044  
 Thorax in acromegaly, iv, 810  
 Threadworm infection, ii, 305  
 Throat in cerebrospinal meningitis, i, 604  
 Thrombi, agglutination of, iv, 527  
     arterial, iv, 537  
     cardiac, iv, 536  
     venous, iv, 537  
 Thrombo-angietitis obliterans, iv, 547

- Thrombophlebitis, idiopathic recurrent, iv, 531  
 of umbilical veins, iv, 546
- Thrombosis, iv, 526  
 in aneurism, iv, 482  
 of aorta, iv, 565  
 apoplexy and, v, 423, 425  
 arterial, iv, 537  
 in cachexia, iv, 530  
 capillary, iv, 538, 548  
 cardiac, iv, 531, 536, 548  
 of carotid artery, iv, 567  
 of cerebral arteries, v, 400  
 in cervical ribs, v, 556  
 in chlorosis, iv, 530  
 of coronary arteries, iv, 569  
   cardiac insufficiency and, iv, 195  
   myocarditis and, iv, 109  
 in dysentery, iv, 530  
 embolism from, iv, 548  
 in enlargement of thymus, iv, 916  
 in gonorrhœa, iv, 529  
 in gout, iv, 531  
 of hepatic artery, iii, 458  
   veins, iv, 545  
 infarction of spleen and, iv, 946  
 of inferior vena cava, iv, 541  
 in influenza, iv, 528  
 in lobar pneumonia, i, 257  
 of lung, ii, 954  
 in malarial fevers, iv, 530  
 of mesenteric arteries, iii, 434  
   hemorrhage and, iii, 307  
   vessels, iv, 542  
 in mitral stenosis, iv, 242  
 in parasitic diseases, iv, 530  
 of peripheral arteries, iv, 568  
 in pneumonia, iv, 528  
 of portal vein, iii, 460; iv, 544  
   ascites and, iii, 659  
 postoperative, iv, 528  
 primary infective, iv, 532  
 pulmonary, iv, 547  
 of renal vessels, iii, 789; iv, 542  
 of retinal arteries, iv, 570  
 in rheumatic fever, iv, 529  
 of spinal cord, v, 256  
 of splenic vein, iv, 948  
 of subclavian artery, iv, 567  
 of superior vena cava, iv, 541  
 in syphilis, ii, 162; iv, 530  
 of thoracic duct, iv, 581  
 in tuberculosis, i, 433; iv, 529  
 in typhoid fever, i, 127, 132; iv, 527  
 in typhus fever, iv, 530  
 venous, iv, 537  
 of vessels of spleen, iv, 546
- Thrombotic ulcers, iii, 350
- Thrush, iii, 50  
 in tuberculosis, i, 431
- Thymic asthma, iv, 918  
 death, iv, 918  
 stridor, iv, 917  
 tracheostenosis, iv, 917
- Thymitis, iv, 928
- Thymus, absence of, iv, 908
- Thymus in acromegaly, iv, 931  
 in acute infections, iv, 934  
 calcification of, iv, 912  
 carcinoma of, iv, 932  
 circulatory disturbances of, iv, 909  
 congestion of, iv, 909, 912  
 convulsions and, v, 794  
 cystic softening of, iv, 912  
 cysts of, iv, 931, 933  
 enlargement of, iv, 912  
 in exophthalmic goitre, iv, 857, 867, 931  
 hemorrhage in, iv, 910  
 hyperplasia of, iv, 912  
 hypertrophy of, iv, 912  
 in hypopituitarism, iv, 817  
 inflammation of, iv, 928  
 in leukemia, iv, 932  
 lipoma of, iv, 932  
 lympho-adenoma of, iv, 932  
 lymphoma of, iv, 932  
 lymphosarcoma of, iv, 932  
 in myasthenia gravis, v, 799  
 œdema of, iv, 909  
 pigmentation of, iv, 912  
 in pseudoleukemia, iv, 932  
 retrograde changes in, iv, 910  
 sarcoma of, iv, 932  
 in sporadic cretinism, iv, 884  
 syphilis of, iv, 930  
 teratoma of, iv, 933  
 tuberculosis of, iv, 929  
 tumors of, iv, 931
- Thyroid gland, achondroplasia and, v, 965  
 in acromegaly, iv, 808  
 actinomycosis of, iv, 830  
 in Addison's disease, iv, 789  
 adenocarcinoma of, iv, 832, 834  
 carcinoma of, iv, 832  
 congestion of, iv, 825  
 diabetes mellitus and, ii, 684  
 echinococcus of, iv, 831  
 in endemic cretinism, iv, 882  
 in exophthalmic goitre, iv, 853, 858  
 in hypopituitarism, iv, 817  
 inflammation of, iv, 825  
 in myxœdema, iv, 890  
 in osteomalacia, v, 948  
 parathyroid glands and, iv, 903  
 sarcoma of, iv, 832  
 scirrhus of, iv, 833  
 syphilis of, iv, 831  
 tuberculosis of, iv, 829  
 tumors of, iv, 832  
 tumors of tongue, iii, 66
- Thyroiditis, iv, 825  
 Graves' disease and, iv, 829  
 infectious diseases and, iv, 826  
 in lobar pneumonia, i, 264  
 myxœdema and, iv, 829  
 sporadic cretinism, and, iv, 883  
 toxic, iv, 826
- Tibial nerve, posterior, paralysis of, v, 559
- Tic convulsif, v, 504, 784  
 douloureux, v, 493
- Tick fever, i, 1020; ii, 133

- Tick infection, ii, 317  
 Tin poisoning, ii, 386  
*Tinea circinata* of mouth, iii, 54  
 Tinnitus aurium, v, 508  
 Toe-drop, ii, 366  
 Tone deafness in aphasia, v, 297  
 Tongue, actinomycosis of, iii, 59  
   anomalies of, iii, 60  
   atrophy of, iii, 66; v, 531  
   black, iii, 61  
   carcinoma of, iii, 66  
   doubling of, iii, 61  
   fibroma of, iii, 66  
   in foot-and-mouth disease, i, 1027  
   frenum of, deformities of, iii, 61  
     ulcer of, iii, 63  
   geographical, iii, 63  
   in hemiplegia, v, 460  
   hyperkeratosis of, iii, 61  
   muscles of, spasm of, v, 532  
   in myxœdema, iv, 891  
   paralysis of, v, 530  
   Riga's disease of, iii, 66  
   in scarlet fever, i, 863, 870  
   spasm of, iii, 62  
   in sprue, iii, 393  
   sucking, iii, 61  
   swallowing, iii, 61  
   syphilis of, ii, 186  
   in tuberculosis, i, 387, 431; iii, 59  
   tumors of, iii, 66  
   wandering rash of, iii, 63  
   worm infection, ii, 323  
 Tonicity of heart muscle, iv, 190  
 Tonsillitis, acute, ii, 848  
   angioneurotic œdema and, iv, 999  
   arthritic deformans and, v, 901  
   chronic, ii, 851  
   enlargement of thymus and, iv, 924  
   exophthalmic goitre and, iv, 850  
   in influenza, i, 539  
   in lobar pneumonia, i, 223  
   nephritis and, iii, 850  
   in purpura rheumatica, iv, 703  
   in rheumatic fever, i, 995  
   tuberculosis and, i, 321  
 Tonsilloliths, ii, 854  
 Tonsils in diphtheria, i, 691, 695  
   diseases of, ii, 847  
   endocarditis and, iv, 152, 154  
   in enlargement of thymus, iv, 916  
   mycosis of, ii, 852  
   pharyngeal, hypertrophy of, ii, 840  
   in scarlet fever, i, 863  
   tuberculosis of, i, 357, 387, 429  
 Tonus of muscles, v, 68  
 Topalgia in hysteria, v, 683  
 Tophi in gout, ii, 744, 746, 751  
 Torticollis, v, 526, 945  
 Touch, sense of, v, 19, 32  
 Tourniquet's sign in scarlet fever, i, 880  
 Toxemia, i, 550  
   apoplexy, and, v, 425  
   in chronic nephritis, iii, 907  
   in diphtheria, i, 563  
   myocarditis and, iv, 124  
   of pregnancy, myocarditis and, iv, 108  
 Toxic dyspnoea, iv, 37  
   myelitis, v, 236  
   purpura, iv, 689  
   thyroiditis, iv, 826  
 Toxins, i, 33, 553  
   epilepsy and, v, 594  
   kidneys and, iii, 773  
   myasthenia gravis and, v, 800  
   nephritis and, iii, 852  
   neuralgia and, v, 726  
 Trachea in aneurism of aorta, iv, 502, 507  
   compression of, iv, 914  
   gumma of, ii, 170  
   obstruction of, ii, 816  
   syphilis of, ii, 170, 906  
   tuberculosis of, ii, 905  
 Tracheal tug in aneurism, iv, 495, 497  
 Tracheitis in measles, i, 906  
 Tracheostenosis, thymic, iv, 917  
 Tracheotomy in diphtheria, i, 722  
 Traction aneurism, iv, 472  
 Transudates in ascites, iii, 659  
 Transverse aorta, aneurism of, iv, 493  
 Traumatic abscess of brain, v, 382  
   arteriovenous aneurism, iv, 510  
   chylothorax, ii, 1059  
   cysts of pancreas, iii, 641  
   hemothorax, ii, 1055  
   hysteria, v, 644  
   lumbago, v, 644  
   mental disturbances, v, 646  
   neurasthenia, v, 643  
   neuroses, v, 642  
   pancreatitis, iii, 612  
   phlebitis, iv, 574  
   psychoses, v, 742  
 Traumatism, abscess of liver and, iii, 497  
   aneurism and, iv, 478  
   angioneurotic œdema and, iv, 1000  
   axillary lymphadenitis and, iv, 735  
   carcinoma of stomach and, iii, 251  
   delirium tremens and, ii, 410  
   dilatation of stomach and, iii, 249  
   erysipelas and, i, 586  
   kidney infections and, iii, 952  
   lymphadenitis simplex and, iv, 730  
   lymphangitis and, iv, 592  
   movable spleen and, iv, 939  
   pleuritis and, ii, 1012  
   rupture of spleen and, iv, 947  
   tetanus and, i, 576  
 Trematode infections, ii, 225  
 Tremor, v, 70  
*Treponema pallidum*, ii, 144  
 Trichina, ii, 295, 297  
*Trichinella spiralis*, ii, 297  
 Trichiniasis, ii, 297  
 Trichinosis, ii, 297  
   typhoid fever and, i, 158  
 Trichiurus, appendicitis and, iii, 360  
 Trichocephaliasis, ii, 295  
 Trichocephalus, ii, 295  
 Trichomonas in carcinoma of stomach, iii, 213  
   intestinalis in gangrene of lung, ii, 988  
 Trichostrongylus, ii, 294  
 Trichuris trichiura, ii, 295

- Tricuspid atresia, congenital, iv, 403  
 insufficiency, iv, 244  
   in tuberculosis, i, 433  
 stenosis, iv, 250  
   in tuberculosis, i, 433  
 valve, safety valve action of, iv, 245
- Trigeminal nerve, diseases of, v, 491  
 paralysis of, v, 493, 494  
   in tabes dorsalis, v, 841  
 neuralgia, v, 733
- Trismus, i, 575; v, 495
- Trombidium tlalsahuate, ii, 323
- Trophic changes in disseminated sclerosis, v, 153  
   in multiple neuritis, v, 584  
 disturbances in general paresis, v, 858  
   in hysteria, v, 685  
   in syringomyelia, v, 224  
   in tabes dorsalis, v, 841  
 functions, anomalies of, v, 79  
 phenomena in tetany, v, 759  
 ulcers, iii, 352
- Tropical abscess of liver, iii, 500  
   big leg, iv, 596  
   chlorosis, ii, 271  
   liver, iii, 494
- Trousseau's phenomena, v, 71, 756
- Truncus arteriosus, persistent, iv, 375
- Trypanosoma brucei, ii, 117  
   evansi, ii, 117  
   gambiense, ii, 119, 121, 122  
   lewisi, ii, 116  
   rhodasiense, ii, 119, 121, 122
- Trypanosomes in blood, ii, 121  
   in cerebrospinal fluid, ii, 121  
   in glands, ii, 121, 127
- Trypanosomiasis, ii, 116  
   human, ii, 119, 127
- Trypsin, iii, 289
- Tsetse-fly disease, ii, 117
- Tubercle bacillus in empyema, ii, 1041  
   in pleuritis, ii, 1011  
   in sputum in tuberculosis, i, 445  
   in tuberculosis of kidney, iii, 984  
 of brain, solitary, v, 319  
   leprosy, i, 528
- Tubercula dolorosa, v, 570
- Tuberculin, i, 304, 307, 505
- Tuberculinic acid, i, 303
- Tuberculoma of brain, encephalic, v, 319
- Tuberculomyces, i, 294
- Tuberculosis, i, 287  
   acute bronchopneumonic, i, 425  
   endocarditis and, iv, 157  
   pneumonic, i, 424  
 of adrenal glands, i, 369; iv, 786, 787  
 amyloid kidney and, iii, 933  
   spleen and, iv, 949  
 aneurism and, iv, 476  
 in animals, i, 293  
 of anus, i, 363  
 of appendix, iii, 385  
 arteriosclerosis and, iv, 454  
 of axillary glands, iv, 744  
 of bladder, i, 366  
 of bloodvessels, i, 360, 386  
 of bones, i, 371
- Tuberculosis of brain, i, 370  
   of breast, i, 368  
   of bronchi, i, 348; ii, 905  
   of bronchial glands, iv, 743  
   bronchopneumonia and, ii, 964  
   of bursæ, i, 373  
   of cervical glands, iv, 743  
     lymphadenitis and, iv, 734  
   chronic nephritis and, iii, 880  
     peritonitis and, iii, 717  
   cirrhosis of liver and, iii, 590, 526  
   congenital, i, 28  
   defect of pericardium and, iv, 345  
   in diabetes mellitus, ii, 706  
   diphtheria and, i, 321, 436  
   of endocardium, i, 360  
   of epididymes, i, 366  
   of epirochlear gland, iv, 744  
   exophthalmic goitre and, iv, 851  
   of Fallopiian tubes, i, 368  
   of fasciæ, i, 373  
   fatty liver and, iii, 467  
   of gall-bladder, i, 364  
   of gall-ducts, i, 364  
   gastric, i, 431  
     ulcer and, iii, 170  
   of genito-urinary tract, i, 365, 389; iii, 1034  
   of gums, iii, 57  
   Hodgkin's disease and, iv, 156, 773  
   influenza and, i, 321  
   of inguinal glands, iv, 744  
   of intestines, i, 361, 432; iii, 387  
   of joints, i, 371  
   of kidneys, i, 365  
   of larynx, i, 348; ii, 876  
   in lead poisoning, ii, 364  
   of lips, iii, 57  
   of liver, i, 363; iii, 541  
   lobar pneumonia and, i, 321, 430  
   of lungs, i, 348  
     pericarditis and, iv, 42  
   of lymph glands, i, 357; iv, 741  
   of lymphatic channels, i, 359  
   tissue, i, 357  
   vessels, iv, 595  
 in lymphoid leukemia, iv, 675  
 malarial fever and, i, 322; ii, 86  
 measles and, i, 907  
 of mediastinal glands, ii, 1111  
 of mesenteric glands, iv, 743  
 miliary, acute, i, 417  
   meningeal, i, 420  
   pulmonary, i, 419  
 of mouth, i, 360; iii, 57  
 multiple neuritis and, v, 515, 586  
 of muscles, i, 373  
 in myeloid leukemia, iv, 670  
 of myocardium, i, 359; iv, 144  
 of nasopharynx, i, 429  
 nephritis and, iii, 850  
 of nose, i, 347  
 of oesophagus, i, 361; iii, 85  
 osteo-arthritis and, v, 982  
 of ovaries, i, 368  
 of pancreas, i, 363; iii, 631  
 of pericardium, i, 359; iv, 66

- Tuberculosis of peritoneum, i, 364; iii, 720  
   of pleura, i, 356  
     pericarditis and, iv, 42  
   pleuritis and, ii, 1006, 1026  
   pneumothorax and, ii, 1073  
   of prostate, i, 366  
   pulmonary, i, 517  
   of rectum, i, 362  
   of salivary glands, i, 363; iii, 70  
   of seminal vesicles, i, 366  
   of skin, i, 373, 386  
   of soft palate, iii, 57  
   of spleen, i, 369  
   of stomach, i, 361, 431  
   syphilis of lung and, ii, 173  
   of tendons, i, 373  
   of testicles, i, 366  
   of thoracic duct, iv, 584  
   of thymus, iv, 929  
   of thyroid glands, i, 369, 435; iv, 829  
   of tongue, iii, 57  
   of tonsil, i, 357, 429  
   tonsillitis and, i, 321  
   of trachea, i, 348; ii, 905  
   trauma and, i, 322  
   typhoid fever and, i, 157, 322  
   of uterus, i, 368  
   of vagina, i, 368  
   whooping cough and, i, 321  
 Tuberculous aortitis, iv, 452  
   bronchopneumonia, i, 425  
   hemoserothorax, ii, 1053  
   lymphadenitis, iv, 741  
   meningitis, i, 420  
     arrhythmia and, iv, 287  
   phlebitis, iv, 576  
   pleuritis, ii, 1010  
     in cirrhosis of liver, iii, 526  
   pneumonia, i, 424  
   syphilide, ii, 166  
   syphiloderm, ii, 184  
   ulcers of intestines, iii, 388  
 Tuberos sclerosus, v, 160  
 Tumor of adrenal glands, iv, 784  
   in appendicitis, iii, 370  
   of appendix, iii, 386  
   of basal ganglia, v, 334  
   of brain, v, 308  
     stem, 335  
   of cavum Meckelii, v, 340  
   of centrum semiovale, v, 276, 334  
   of cerebellum, v, 338  
   in cirrhosis of stomach, iii, 277  
   of corpora quadrigemina, v, 336  
   of corpus callosum, v, 276, 335  
   of crura, v, 335  
   in cyst of pancreas, iii, 643  
   extracerebellar, v, 338  
   of fourth ventricle, v, 338  
   of frontal lobe, v, 331  
   in gangrenous pancreatitis, iii, 618  
   hydrocephalus and, v, 356  
   in hypertrophic stenosis of pylorus, iii, 271  
   in hypopituitarism, iv, 817  
   in inflammation of spleen, iv, 942  
   of intestines, iii, 424, 433  
 Tumor, intracerebellar, v, 338  
   of kidney, iii, 994, 1003, 1005  
   of lips, iii, 45  
   of liver, iii, 541, 546  
   of lung, ii, 997  
   of lymph nodes, iv, 747, 752  
   of mediastinum, ii, 1093  
   of medulla oblongata, v, 337  
   of mouth, iii, 52  
   of nerves, v, 568  
   of paracentral convolutions, v, 332  
   of parathyroid glands, iv, 904  
   of parietal lobe, v, 333  
   of pelvis of kidney, iii, 1006  
   of pericardium, iv, 92  
   of peritoneum, iii, 730, 740  
   phantom, iii, 299  
   of pineal gland, v, 336  
   of pituitary body, iv, 804  
   of pleura, ii, 1060, 1064  
   of pons, v, 336  
   of quadrigeminal bodies, v, 282  
   of salivary glands, iii, 72  
   of spinal cord, v, 196  
   of spleen, iv, 950  
   in suppurative pancreatitis, iii, 624  
   of tegmentum, v, 336  
   of temporal lobe, v, 331  
   of thoracic duct, iv, 587  
   of thymus, iv, 931  
   of thyroid gland, iv, 832  
   of tongue, iii, 66  
 Tunnel anemia, ii, 271  
 Twilight states, v, 44  
 Twitchings, fibrillary, v, 70  
 Tydeus molestus, ii, 323  
 Tympanites, iii, 298  
   in acute peritonitis, iii, 681  
 Typhilitis, iii, 355  
 Typhoid bacillus, acute cholecystitis and, iii, 561  
   in blood, i, 78  
   endocarditis, iv, 161  
   fever, i, 67  
     alcoholism and, i, 159  
     ambulatory, i, 153  
     amebic dysentery and, i, 158  
     apoplexy and, v, 417, 425  
     appendicitis and, i, 159  
     arteriosclerosis and, iv, 454  
     Asiatic cholera and, i, 158  
     bacillary dysentery and, i, 777  
     bacterial infections and, i, 159  
     cerebral thrombosis and, v, 400  
     cirrhosis of liver and, iii, 509  
     diphtheria and, i, 158  
     disseminated sclerosis and, v, 141  
     erysipelas and, i, 158  
     in fetus, i, 156  
     gonorrhoea and, i, 158  
     hemorrhagic, i, 153  
     in infancy, i, 154  
     inflammation of spleen and, iv, 941  
     influenza and, i, 158  
     intussusception and, i, 159

- Typhoid fever, lobar pneumonia and, i, 122, 207, 267  
 malarial fever and, i, 156  
 malignant, i, 152  
 mild, i, 152  
 multiple neuritis and, v, 575  
 myocarditis and, iv, 108, 117  
 nephritis and, iii, 849  
 neurasthenia and, v, 629  
 scarlet fever and, i, 158  
 thrombosis in, iv, 527, 530  
 tuberculosis and, i, 157, 322  
 typhus fever and, i, 158  
 form of influenza, i, 544  
 pellagra, ii, 482  
 spine, i, 147  
 Typhus fever, i, 924  
   typhoid fever and, i, 158  
 Tyroglyphus, ii, 323  
 Tyrosin in urine, iii, 816  
 Tyrotoxin, ii, 467  
 Tyrotoxisms, ii, 467
- U**
- ULCER, duodenal, iii, 191, 344  
   subphrenic abscess and, iii, 701  
   pyopneumothorax and, iii, 710  
 of frenum of tongue, iii, 63  
 gastric, iii, 168  
   chronic peritonitis and, iii, 714  
   intestinal hemorrhage and, iii, 307  
   subphrenic abscess and, iii, 701  
   pyopneumothorax and, iii, 710  
   in tuberculosis, i, 432  
 of intestine, iii, 343  
 of cesophagus, iii, 84  
 peptic, iii, 168  
   jejunal, iii, 349  
 of rectum in tuberculosis, i, 432  
 of stomach, arteriosclerosis and, iv, 464  
   hematemesis from, iii, 283  
   hyperacidity and, iii, 114  
   syphilitic, ii, 174  
 trophic, iii, 352  
 in typhoid fever, i, 101  
 uremic, in chronic nephritis, iii, 899  
 Ulcerated syphilide, ii, 166  
 Ulcerative cholecystitis, iii, 563  
   endocarditis, iv, 148, 151  
   stomatitis, iii, 49  
   syphiloderm, ii, 185  
 Ulnar nerve, paralysis of, v, 549  
 Umbilical veins, thrombophlebitis of, iv, 546  
 Uncinaria americana, ii, 270  
   craniformis, ii, 271  
   duodenalis, cirrhosis of liver and, iii, 510  
   vulpis, ii, 271  
 Uncinariasis, ii, 271  
 Uncinate fits, v, 332  
 Undulant fever, i, 664  
 Unilateral paralysis, v, 108  
 Urachial cysts of peritoneum, iii, 739  
 Uranium, rays from, ii, 339  
 Urea, intoxication and, ii, 527, 534  
   in protein metabolism, ii, 586  
 Uremia, ii, 534; iii, 830  
   acute, iii, 841  
   auto-intoxication and, ii, 534  
   chronic, iii, 842  
   in gout, ii, 750  
   in mercury poisoning, ii, 380  
 Uremic ulcers in chronic nephritis, iii, 899  
   of intestines, iii, 350, 838  
   of cesophagus, iii, 84  
 Ureter, inflammation of, iii, 974  
   in pyelitis, iii, 954  
   in renal calculus, iii, 1017  
   stricture of, kidney infections and, iii, 952  
   in tuberculosis of kidney, iii, 980  
 Ureteral catheterization, iii, 939  
 Urethritis, iii, 974  
   kidney infections and, iii, 952  
 Urethra, stricture of, kidney infections and, iii, 952  
 Urethritis, tuberculosis of kidney and, iii, 978  
   in typhoid fever, i, 144  
 Uric acid in blood, ii, 539, 736  
   calculus, iii, 1014  
   in diabetes mellitus, ii, 703  
   excretion of, in gout, ii, 737  
   metabolism in gout, ii, 732  
   purins and, ii, 733  
 Urinary excretion, anomalies of, iii, 791  
   lithiasis, iii, 1007  
   tract in typhoid fever, i, 86  
 Urine in abscess of liver, iii, 592  
   of kidney, iii, 959  
   in acromegaly, iv, 812  
   in acute catarrhal cholangitis, iii, 553  
   dilatation of stomach, iii, 252  
   hepatitis, iii, 495  
   nephritis, iii, 861  
   peritonitis, iii, 684  
   yellow atrophy of liver, iii, 476  
   in Addison's disease, iv, 795  
   air in, iii, 806  
   albumin in, iii, 794  
   ammoniacal, iii, 793  
   in amyloid kidney, iii, 935  
   in appendicitis, iii, 371  
   in arteriosclerosis, iv, 463  
   in arthritis deformans, v, 916  
   bile in, iii, 828  
   in blackwater fever, ii, 114  
   blood in, iii, 820  
   in carcinoma of stomach, iii, 207  
   in cardiac insufficiency, iv, 210  
   casts in, iii, 798  
   in chlorosis, iv, 651  
   in chronic nephritis, iii, 882, 900  
   perihepatitis, iii, 492  
   chyle in, iii, 805  
   in cirrhosis of liver, iii, 526, 539  
   in congestion of liver, iii, 479, 485  
   creatinin in, iii, 813



- Urine, cryosecopy of, iii, 792  
 crystals in, iii, 800  
 cylindroids in, iii, 800  
 cystin in, iii, 817  
 in diabetes mellitus, ii, 700  
 in dilatation of stomach, iii, 252, 257  
 in erysipelas, i, 588  
 fat in, iii, 827  
 in fat necrosis, iii, 609  
 fibrin in, iii, 806  
 in gastric ulcer, iii, 176  
 glucose in, iii, 821  
 glycuronic acid in, iii, 823  
 in gout, ii, 748, 749  
 hemogentisic acid in, iii, 824  
 in hyperacidity, iii, 117  
 in hypernephroma, iii, 997  
 in jaundice, iii, 453  
 in kala-azar, iv, 972  
 in kidney of pregnancy, iii, 868  
 lactose in, iii, 822  
 leucin in, iii, 816  
 levulose in, iii, 821  
 in lobar pneumonia, i, 232  
 in lymphoid leukemia, iv, 672  
 in malarial fevers, ii, 67  
 in measles, i, 905  
 in migraine, v, 718  
 in mitral insufficiency, iv, 236  
 in myasthenia gravis, v, 805  
 in myeloid leukemia, iv, 666  
 in necrosis of pancreas, iii, 617  
 in nephritis, iii, 959  
 in nephroptosis, iii, 756  
 in neurasthenia, v, 632  
 in osteomalacia, v, 952  
 in paraneuritis, iii, 972  
 in pellagra, ii, 484  
 pentoses in, iii, 822  
 in periodic paralysis, v, 811  
 in pernicious anemia, iv, 635  
 in plague, i, 629  
 proteins in, iii, 794, 818  
 pus in, iii, 805  
 in pyelitis, iii, 958  
 in pyelonephritis, iii, 958  
 in pyogenic infections of kidney, iii, 958  
 in pyonephrosis, iii, 959  
 in Raynaud's disease, iv, 989  
 in serofibrinous pleuritis, ii, 1017  
 spermatozoa in, 800  
 sugars in, 821  
 in tuberculosis, i, 389  
     of kidney, iii, 983  
     of peritoneum, iii, 726  
 in typhoid fever, i, 174  
 tyrosin in, iii, 816  
 in uremia, iii, 830  
 in whooping cough, i, 659
- Urobilinuria, iii, 810  
 in jaundice, iii, 454
- Uro-erythrinuria, iii, 810
- Urogenital system, nematodes of, ii, 315
- Urostealiths, iii, 1009
- Urticaria, giant, iv, 998  
 in Henoch's purpura, iv, 709
- Urticaria of mouth, iii, 53  
 œdematosa, iv, 998  
 purpura and, iv, 704  
     rheumatica and, iv, 703  
 in Raynaud's disease, iv, 990  
 in syringomyelia, v, 224  
 in typhoid fever, i, 101
- Uterus, diphtheria of, i, 699  
 perforation of, acute diffuse peritonitis and, iii, 673  
 tuberculosis of, i, 368

## V

- VACCINATION, i, 822  
 accidental, i, 841  
 in purulent pleuritis, ii, 1048  
 in smallpox, i, 843  
 in tuberculosis, i, 436, 844
- Vaccine, nephritis and, iii, 849  
 treatment of typhoid fever, i, 187  
 virus, i, 828
- Vaccines, tuberculosis, i, 305
- Vaccinia gangranosa, i, 842  
 generalized, i, 841
- Vagal attacks, v, 523
- Vagina, tuberculosis of, i, 368
- Vagus nerve, aneurism and, iv, 501  
 arrhythmia and, iv, 288  
 bradycardia and, iv, 282, 284  
 paralysis of, v, 517  
 Stokes-Adams syndrome and, iv, 307  
 tachycardia and, iv, 276, 280, 304
- Valleix's points, v, 17, 727, 734, 737
- Valsalva, sinus of, aneurism of, iv, 492
- Valves of heart, aneurism of, iv, 482  
 combined lesions of, iv, 259  
 rupture of, aortic insufficiency and, iv, 219, 227
- Valvula foraminis ovalis, iv, 326
- Valvular lesions, hypertrophy of heart from, iv, 178  
 myocarditis and, iv, 128  
 in septic endocarditis, iv, 163  
 pneumothorax, ii, 1083
- Varicella, i, 849
- Varices in stomach, hematemeses from, iii, 283
- Varicose aneurism, iv, 510  
 groin glands, ii, 308
- Variola, i, 783  
 myocarditis and, iv, 108
- Varioloid, i, 809
- Vascular changes in disseminated sclerosis, v, 153  
 crises, iv, 460  
 goitre, iv, 839  
 lesions in lobar pneumonia, i, 257  
 stasis, iv, 20
- Vasomotor and trophic disorders, iv, 975
- Vegetable poisoning, ii, 468
- Vegetative aortitis, acute, iv, 456  
 endocarditis, iv, 151
- Veins in aortic insufficiency, iv, 224  
 portal, in splenic anemia, iv, 955

- Veins in typhoid fever, i, 128  
 Vena cava, inferior, thrombosis of, iv, 541  
   left superior, persistent, iv, 439  
   right superior, absence of, iv, 440  
   superior, aneurism and, iv, 504  
   thrombosis of, iv, 541  
 Venal distomatosis, ii, 239  
 Venous embolism, iv, 552  
   stasis, congenital cyanosis and, iv, 334  
   hematemesis from, iii, 283  
   in mitral insufficiency, iv, 236  
   in pericarditis, iv, 21, 48  
   thrombosis, iv, 537  
     peripheral, iv, 539  
 Ventricles, fibrillation of, iv, 106  
   left, hypertrophy of, iv, 185  
   right, hypertrophy of, iv, 185  
   standstill of, iv, 89  
 Ventricular asystole, iv, 89  
 Verrucose endocarditis, iv, 150  
 Vertigo, v, 30, 511, 515  
 Vesicular emphysema of lungs, ii, 982  
   eruption in syphilis, ii, 165  
 Vestibular nerve, diseases of, v, 511  
 Vibration sense, v, 18  
 Villous arthritis, v, 929  
 Virulence, bacterial, i, 37  
 Virus, filterable, i, 19  
   in carlet fever, i, 860  
   ultramicroscopic, i, 19  
 Visceral lesions in syphilis, ii, 169, 190  
 Visceroptosis, iii, 747  
 Vision, v, 25, 480  
 Visual aphasia, v, 298  
 Vitamine, ii, 647  
   in scurvy, ii, 799, 805  
 Vitiligo in exophthalmic goitre, iv, 866  
 Vocal cords, paralysis of, in tuberculosis  
   of bronchial glands, iv, 743  
   fremitus in lobar pneumonia, i, 246  
   in tuberculosis, i, 407  
 Voit's theory of protein metabolism, ii, 592  
 Volvulus, iii, 412  
   of omentum, iii, 418  
 Vomiting, juvenile, iii, 145  
   nervous, iii, 142  
   periodical, iii, 144  
   reflex, iii, 145
- W**
- WANDERING rash of tongue, iii, 63  
   spleen, iv, 938  
 Wassermann reaction in cerebrospinal  
   syphilis, v, 826, 827  
   in general paresis, v, 851  
   in syphilis, ii, 191  
   in tabes dorsalis, v, 847  
 Watsonius watsoni, ii, 239  
 Waxy casts, iii, 799  
 Weber's syndrome, v, 369  
   test, v, 24  
 Weil's disease, i, 1014  
 Weir Mitchell's disease, iv, 1025  
 Werdnig-Hoffmann type of spinal muscu-  
   lar atrophy, v, 87
- Wernicke's aphasia, v, 299  
   disease, v, 365  
   zone, v, 300  
 Wet brain, ii, 415  
 Whipworm infection, ii, 295  
 "White line" in Addison's disease, iv, 792  
 Whooping cough, i, 656  
   apoplexy and, v, 417  
   diphtheria and, i, 659  
   scarlet fever and, i, 659  
   tuberculosis and, i, 321  
 Widal reaction, i, 162  
 Wood alcohol poisoning, ii, 298  
 Wool-sorter's disease, i, 639  
 Word amnesia, v, 76  
   blindness, v, 54, 76, 298  
   deafness, v, 54, 75, 297  
 Wounds of heart, insufficiency and, iv, 191  
 Wright's stain, ii, 91  
 Wrist-drop, ii, 366  
 Writer's cramp, v, 744  
 Writing, anomalies of, v, 74  
 Wryneck, v, 526
- X**
- XANTHELASMA in jaundice, iii, 452  
 Xanthin calculus, iii, 1009  
 Xanthoma in diabetes mellitus, ii, 705  
 Xerostomia, iii, 68  
 X-rays, ii, 335  
   alopecia from, ii, 336  
   atrophy of skin from, ii, 336  
   burns, ii, 336  
   cancer from, ii, 336  
   in carcinoma of stomach, iii, 215  
   in congenital diseases of heart, iv, 444  
   dermatitis from, ii, 335  
   in diagnosis of acromegaly, iv, 814  
   of cholelithiasis, iii, 585  
   in enlargement of thymus, iv, 924  
   eye lesions from, ii, 336  
   gangrene from, ii, 336  
   in gastric ulcer, iii, 181  
   in lobar pneumonia, i, 247  
   nephritis and, iii, 853  
   in spondylitis deformans, v, 927  
   sterility and, ii, 337  
   swelling of muscles from, ii, 336
- Y**
- YELLOW fever, i, 936  
   nephritis and, iii, 849
- Z**
- ZINC poisoning, ii, 386  
 Zoöparasitic diseases, ii, 217







# Date Due

Amey



1041

Isler

Modern Medicine

QsF

1015

W. F.

copy 1

